

An Introduction To **CLINICAL NEUROLOGY**

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By

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With A Foreword By

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“DEVYANI”

**And we, who are left, must grow old with the years,
knowing failure and heartache and blame,
while you stand on the hill with the wind in your hair,
Young and beautiful, always the Same!**

FOREWORD

Of all the specialities in medicine, neurology is perhaps the one in which clinical diagnosis and management must be based firmly upon a sound knowledge of the anatomy, physiology and pathophysiology of the central and peripheral nervous system, including the neuromuscular apparatus. The advent of new technologies, and above all the introduction of computerised tomography, has transformed the diagnosis and management of diseases of the nervous system. This and other investigative techniques must be employed judiciously and according to a logical plan of investigation based upon the sound and detailed analysis of the clinical manifestations in any individual case. There are indeed many neurological disorders in which diagnosis rests upon a careful clinical history and in which even physical examination may be uninformative. Migraine is a good example. Thus to possess and to exploit appropriately even the most sophisticated modern investigative methods without establishing first a basis of sound clinical diagnostic skill is counter-productive. In other words, the neurologist and the general physician wishing to deal satisfactorily with patients suffering from neurological disease requires that basic understanding of neurological pathophysiology without which the practice of this specialty is quite unsatisfactory.

Dr. Anil Desai and Dr. Bindu Desai have done their Indian colleagues a service in writing this short and succinct commentary upon the principles of clinical neurology. The Bombay school has been outstanding in the development of neurological thought and practice in the Indian sub-continent. I am sure that the medical students, family physicians and young doctors training in internal medicine and neurology who read this book will find that if they follow its principles and precepts, it will enliven their practice and promote a clearer understanding of the function of the nervous system in health and disease.

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PREFACE

Neurology did not enjoy the status of a separate subspeciality in India till the early fifties. General physicians taught neurology at both undergraduate and postgraduate levels. Even today, undergraduate students are not assigned rotations in neurology wards. Moreover, neurology is still not established as a speciality in many Indian medical colleges. We have been aware of the need for a textbook of neurology that would be useful to undergraduates and family physicians. The senior author has often found postgraduate students in Medicine lacking a sound knowledge of the **fundamentals** of neurology, and carrying very vague concepts of common problems in clinical neurology.

This book introduces the subject of clinical neurology in a concise and lucid manner. There has been no attempt to include rare disease entities and references. However, in each chapter detailed classification of diseases have been included for students desirous of further study. The aim has been to cover common problems in neurology as they apply to the Indian subcontinent. Modern concepts of pain, migraine, and **classification of seizures are emphasised**. The eager student should be stimulated to read in depth, and some may even be drawn to a career in neurology. The first two chapters deal with signs and symptoms. Certain clinical conditions such as migraine, coma, cerebro-vascular disease etc., are dealt with in depth in the initial chapters for the basic understanding and stressing the importance of good history taking and detailed examination. Later chapters describe specific clinical conditions in a **crisp and simple manner**. The authors have tried to limit the size of the book, so that it is inexpensive. Regretably, this has led to fewer illustrations being included.

We would like to acknowledge the help rendered by Dr. N.H. Wadia and by many undergraduate medical students who gave us many useful suggestions. Special thanks are due to Mr. Ashit Parekh who willingly acceded to our request to draw all the illustrations, and to Mrs. S.S. Patwardhan for the secretarial help.

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CHAPTER - I

HISTORY

“Of Charcot it may be said with truth that he entered neurology in its infancy and left it at its coming of age, largely nourished by his own contributions”.

I.S. Wechsler.

Clinical neurology flowered in the nineteenth century with two major centers in Europe dominating the field. The Hospital of the Saltpetriere in Paris was converted from an asylum for infirm and abandoned old women into a world renowned center for research and training. Here Jean Martin Charcot (1825-1893) worked with enormous energy and an iron will founding a clinical neurology service that included Pierre Marie and Babinski amongst its students. The National Hospital for Nervous Diseases in London rivalled the Saltpetriere in its prestige and distinguished faculty. John Hughlings Jackson (1835-1911) and Sir William Gowers (1845-1915) laid the foundations of clinical neurology, their writings providing excellent reading even today. In the middle of the last century the technique of the neurological examination was gradually developed.

The art of history taking, always important in clinical medicine is crucial to neurology. A proper chronological account of the patients symptoms aid in the diagnosis. In India, the clinician has to deal with additional special problems. Illiteracy abounds, superstition is rampant and irrelevant problems such as constipation are given undue importance. India is a multilingual nation and very often the physician and the patient do not share a common language. Moreover, multiple systems of medicine are being practised. Much patience and practice are required before a physician can obtain a history where not only is the ‘chaff separated from the grain’ but a proper time frame constructed which allows a tentative diagnosis to be reached. For instance, a young man complains of progressive weakness of the right arm and leg. When questioned for other symptoms he mentions headaches but immediately adds that he had attributed them to ‘overwork’ and poor eyesight. We

are now aware that he also has headaches and poor vision; further inquiry reveals the headache **preceded** the visual problems by several months. The headaches began six months ago and **were increasing** in severity. Worse in the morning, increased by coughing or straining at stools, the headache was localized to the left frontal region. Initially intermittent, it later became constant and was not relieved with analgesics. The visual problem began a month ago and was characterized as a difficulty in focusing objects with blurring of distant images. On the basis of this history we can suspect that the patient's problems are caused by a slow growing mass located in the left frontal region which has resulted in intra-cranial pressure, papilloedema and focal neurologic deficit. Examination revealed early papilloedema and a mild right hemiparesis with increased tone and deep tendon reflexes. At surgery he was found to have a glioma.

The above case illustrates the importance of certain symptoms in neurologic practice. Headaches, dizziness, loss of consciousness, limb weakness and numbness are but a few of the common complaints. **One has to know chronological order in which the events occurred, to get an insight into the clinical course, whether changes were sudden or gradual** in onset and if the symptom has progressed or stabilized. We will now consider some of the symptoms commonly encountered in neurology, examine their pathophysiology and describe their characteristics.

PAIN "But pain is perfect misery, the worst of evils, and, excessive, overturns All Patience."

John Milton. *Paradise Lost*. Book Six. 456-464.

Pain should be described in terms of its **location**, duration and character. Are there any precipitating or relieving factors? Does the pain tend to occur at any particular time of the day? Research into the basic mechanisms of pain has expanded rapidly in the past few years. There have been exciting new discoveries showing that the brain synthesises its own "morphine" like substance-enkephalins and endorphins. Pain transmission in the central nervous system is also linked to neurotransmitters such as serotonin. Both these systems controlling the production of pain are presently being investigated for therapeutic applications. Already serotonin uptake blocking agents such as amitriptyline have been shown to ameliorate migraine. **Table 1.1 lists the pain sensitive structures in the cranium.**

TABLE 1.1

PAIN SENSITIVE STRUCTURES.

Cranial and Extracranial

1. Coverings of the skull.
2. The skull and the periosteum.
3. Orbital contents.
4. Accessory nasal sinuses.
5. Teeth and ears.

Intracranial

1. Pia covering the circle of Willis and their major branches.
2. Parietal dura along the base.
3. Large venous sinuses.
4. Falx and tentorium.

Nerves

Above tentorium : Trigeminal

Below tentorium : Glossopharyngeal and vagus and cervical roots.

Note that the brain itself is not sensitive to pain, but, pain occurs when any of the structures listed above are either inflamed, stretched or dilated. The upper cervical roots supply the dura around the foramen magnum. The *Character* of pain varies according to cause. Pain due to inflamed dura is usually dull aching whilst pain secondary to dilatation of blood vessels is throbbing in nature. In migraine, hypertension, vascular tumours, post lumbar puncture, infective fevers and drug withdrawal, the headaches are throbbing in character, and are either temporal or occipital. Eyestrain, sinusitis and emotional tension cause tight bandlike sensations whereas the pain of meningitis, subarachnoid haemorrhage is described as steady and severe. Neuralgic pain is shooting in character. Headaches may be generalised or localized depending on the site of pain. The pain is referred to fronto-temporal regions in supratentorial tumours. Posterior auricular and occipital pain occurs in infratentorial lesions and meningitis, whereas the pain is referred to the vertex in sellar and parasellar lesions. The *intensity* of pain varies with the cause. It is mild in diseases of the eye with exception of glaucoma, moderate in tension headaches or tumours and severe in meningitis, subarachnoid haemorrhage and migraine. *Psychogenic headaches* are usually described with extravagant epithets such as

'burning pain' 'inside feels as though it will burst' or so severe that the patient wants to bang his head against the wall. They may be either hysterical or tension headaches. The latter are precipitated by anxiety, fatigue and apprehension while the hysterical headaches are the result of personality disorder. The hysterical headaches are present throughout the day and night and are of many years duration. The tension headaches are worst in the evening and occur only during the duration of tension.

The pain of *sinusitis* is generalized but can be referred to the vertex if the sphenoid sinus is involved. Dull aching, of mild to moderate intensity, it may be relieved by lying down with diseased side up. When the frontal sinuses are involved pain occurs in the early morning and decreases as the day wears on. It is aggravated by bending down and emotional tension and is relieved by analgesics.

Headaches caused by tumours are steady, dull aching and are worse in the early morning hours. They are aggravated by coughing, stooping or straining. Of moderate intensity they are intermittent and do not interfere with sleep. *Headache due to meningitis* is present throughout the day, severe throbbing pain of a generalised nature associated with stiffness of the neck occurs.

MIGRAINE headaches are periodic and the interval between two **attacks being variable**. They may occur as often as twice a week or once every few months. They start early in life, and no social, intellectual or economical group is immune to them. Almost always a history of migraine is present in the family. The headache of *common migraine* is unilateral at the onset becoming generalized as the headache progresses. The side on which the pain occurs may vary in each episode.

Severe **throbbing** pain occurs associated with photophobia, nausea and vomiting. The patient likes to be left alone in a quiet dark room. Vegetative symptoms such as nausea, vomiting, fatigue, chills, thirst and oliguria may occur. There is no prodrome in common migraine. The mechanism of migraine is attributed to a neurovascular reaction with diminution of cerebral blood flow during the prodrome and increased cerebral and extracranial blood flow during the headache. The pain of migraine is caused by dilatation of the large extracranial arteries with a release of a bradykinin like substance around the affected vessels. Plasma serotonin content increases before a migraine headache and

then falls during the headache phase in most patients.

Other types of migraine include :

- 1) **PREMONITARY MIGRAINE** episodes in which mood changes, **thirst, craving for sweet foods and drowsiness precedes the headache** by twenty-four hours.
- 2) **PRODROMAL**
 - a. **CLASSIC MIGRAINE** visual disturbances occur such as teichopsia, fortification spectra and field defects. The visual scotomas may have various shapes and are often bordered by moving streaks of light. They are sometimes described as jagged or as resembling the ramparts of a medieval castle (fortification spectra). Other focal neurological symptoms such as hemiparesis or dysphasia may also occur. These symptoms precede the headache by ten to sixty minutes. Retinal migraine is a subcategory of classic migraine in which the vasoconstriction is limited to the branches of one ophthalmic artery.
 - b. **PROTRACTED PRODROMAL MIGRAINE** episodes in which focal neurologic symptoms persist into the headache phase. This type includes most cases of basilar and hemiplegic migraine.
- 3) **MIGRAINE EQUIVALENT** The gradual onset and subsidence of focal neurological symptoms similar to a migrainous prodrome but where no headache occurs.
- 4) **COMPLICATED MIGRAINE** retinal or neurological deficit persists after the usual duration of migraine headache or migraine equivalent.
- 5) **CLUSTER HEADACHE** also known as migrainous neuralgia and Horton's histamine cephalgia, this disorder occurs predominantly in males. The headaches occur in clusters, recurring in bouts that last from weeks to months. Characteristically waking the patient in the early hours of the morning the headache is associated with severe retro-orbital pain, redness and tearing in one eye and nasal stuffiness.

Management Individual attack of migraine can either be relieved by administration of an analgesic like aspirin, paracetamol or analgin, or Ergotamine tartrate orally in the dose of 1 to 2 mg at the onset of the headache. If no relief the dose can be repeated every ½ hour till 3 to 5

tablets are given. Ergot preparations should be avoided in **peripheral vascular disease**, coronary disease, pregnancy and thyrotoxicosis.

For prevention of recurrent attacks amitryptiline in the dose of 10 to 25 mgm at bed time has been found to be most useful. Methysergide and pizotifen are recent introductions for the prevention but are not available in the country. Clonidine, propranolol and anticonvulsants have also been tried for prevention of attacks.

Hypertensive Headache is said to be occipital in location. It is important to remember that headache is as common in hypertensives as in normotensives. Many hypertensives never have headaches and 'hypertension headache' has no specific diagnostic feature.

Severe unrelenting headache of sudden onset in a patient otherwise previously well should lead one to suspect *subarachnoid haemorrhage* secondary to an aneurysmal rupture. If the bleed is large patient **suddenly feels something snapping in the head, gets a bursting headache** accompanied by vomiting, convulsions, coma or focal neurological deficit. If the bleed is minimal, the headaches may be ignored both by patient and the doctor.

Almost 80 percent of aneurysms are situated in the anterior half of circle of Willis and the **rupture** will produce frontal headaches. While occipital and nuchal pain is produced by bleed from the posteriorly situated aneurysms. Onset of sudden headaches with signs of meningeal irritation should alert the examiner to the possibility of a subarachnoid bleed.

Headache following *lumbar puncture* and *convulsions* or due to a *hangover* is either deep dull aching or throbbing and is generalized. It increases by body effort, **exertion or movement of the head**.

Vomiting secondary to intracranial causes such as tumour or meningitis is sudden, spasmodic, not associated with nausea and projectile in nature.

A variety of *visuat disturbances* occur in neurologic disease. Dimunition of vision occurs in the form of dark spots which may be small or large, single or multiple and are known as scotoma. Quadrantic or hemianopic *fiel'd defects* of gradual onset do not usually cause the patient to complain of visual symptoms. Sometimes a history of a

tendency to bump into objects on the affected side can be elicited. Rarely blindness in one eye goes unnoticed by the patient. Visual loss due to papilloedema begins with blurring of vision for distance objects. Total loss of vision occurs late in the course of the disease. Patients with well developed papilloedema may have very brief obscurations of vision: "grey outs" or "black outs", dimming of vision usually involving one eye at a time for a few seconds and clearing completely. Visual loss in papillitis or retrobulbar neuritis is complete in a few hours to a day. **Patients complain of pain on moving the eyes.** Perimetric examination shows an enlarged blind spot in papilloedema with greater constriction of vision to the colour red than to white. In retrobulbar neuritis there is a central scotoma and the constriction of vision is equal to all colours.

Diplopia or double vision is due to acquired weakness of one or more of the extraocular muscles. Monocular diplopia is rare and results from disorders of the crystalline lens or a detachment of the retina. Whenever a patient complains of double vision the following questions must be asked: Are the images side by side, one on top of the other or a combination of the two? If the images are side by side the lateral or medial rectus muscle is involved whereas if the images are one on top of the other either the obliques or the superior or inferior rectus must be affected. There are two main laws that govern diplopia: a) the separation of images is greatest when the eyes are moved in the normal direction of action of the affected muscle. b) The false image is the farthest image.

Ptosis or drooping of the eyelid occurs when the pupillary margin is obscured by the lid. Ptosis may be unilateral or bilateral. Its causes are legion. Normal eyelid tone is maintained by two muscles: a) Mueller's muscle or the superior tarsalis which is innervated by the sympathetic chain. b) The levator palpebrae superioris innervated by the oculomotor. Therefore ptosis can result from a lesion anywhere along the course of the oculosympathetic pathway which begins in the hypothalamus, descends to the thoracic spinal cord and then ascends via the sympathetic chain and the carotid artery to supply the globe. Ptosis can also result from a lesion of the mid-brain nucleus of the levator and along the course of the oculomotor nerve. In Marcus-Gunn phenomenon the eye closure occurs with chewing movement and the patient appears to be winking while eating. The inverted Marcus-Gunn phenomenon occurs in recovering facial palsy where in with opening of the mouth the lid droops. The causes of Ptosis are listed in Table 1.2.

TABLE 1.2
CAUSES OF PTOSIS

Lesions in the muscle :	Ocular myopathy
	: Myasthenia gravis
	: Myotonic dystrophy
Lesions of the eyelid :	Lid or orbital tumour (Haemangioma, dermoid)
	Eyelid infection
Lesions of the	
sympathetic :	Horner's syndrome due to—Syringomyelia
	Cervical rib
	Pancoast tumour
	Cavernous sinus lesion
	Tabes
	Raeder's syndrome
	Cervical cord tumours.
Lesions of the oculo	
motor nerve :	Vascular occlusion - Benedict's or Weber's syndromes
	Basal meningitis - Pyogenic, tuberculous etc.
	Aneurysms of the internal carotid and posterior cerebral arteries
	Diabetes
	Polyneuritis cranialis
	Increased intracranial pressure - subdural haematoma
	Sphenoid ridge meningioma
	Pituitary apoplexy

Inability to close the eyes is secondary to weakness of the orbicularis oculi and lesions of the facial nerve. Weakness of eye opening and closure together is almost always due to muscle pathology. The third and seventh nerves are rarely affected together as in mucormycosis of the orbit and polyneuritis cranialis.

Difficulty in phonation may be characterized as slurred speech, nasal twang, hoarseness of voice or total aphonia. *Slurred speech* is usually due to lesions involving the facial muscles either of the upper or lower motor neurone. Slurred speech can also be due to cerebellar

pathology. *Nasal voice* may be accompanied by nasal regurgitation and is due to unilateral or bilateral palatal weakness. Lesions of the recurrent laryngeal branch of the vagus nerve result in *hoarseness of voice*. Total paralysis of both vagus nerves is not compatible with life.

Dysphagia or difficulty in swallowing results from weakness of the pharyngeal muscles. It may be due to lesions of the upper motor neuron as in 'psuedobulbar palsy', or of the lower motor neuron as in **poliomyelitis**, or of the motor end plate as in myasthenia.

Dizziness is a common complaint which is employed by the patient for a variety of symptoms. It is essential that the examiner clarifies at the outset what the patient means when he complains of being dizzy. **Vertigo** is an illusion of movement which results from defects either in the peripheral or the central nervous system. Ill defined lightheadness is not to be confused with vertigo. In vertigo the patient feels either that he is moving around or that objects are rotating around him. Some authors define the former as subjective vertigo and the latter as objective vertigo but this division has little diagnostic value. Once you have determined that the patient indeed has true vertigo, it is necessary to inquire into related problems such as pain or discharge from the ear, deafness or ringing of the ears, all suggesting middle or inner ear pathology. Numbness of the face or body, double vision, clumsiness of movement, indicate vertebro-basilar ischaemia. Epileptic vertigo is rare and is followed by a period of impaired responsiveness. Postural vertigo is brought on by change of posture and is generally secondary to labyrinthine disorders. Hyperventilation attacks are a common cause of vertigo especially in tense individuals. In Meniere's disease the vertigo is periodic, occurring at irregular intervals, individual attack may last few hours to few days. It is common after the age of 30 years and affects females more than males. Tinnitus and deafness occurs later in the course of the disease. Vomiting or severe nausea is usually associated. Sweating, pallor, drop in blood pressure and even syncope can occur during the attack. It is thought to be due to hydrops of the membranous labyrinth. The acute attack is managed by making the patient lie down and find a neutral position of the head in which the vertigo stops. Prochlorperazine in the dose of 10 to 25 mgms. may be injected intramuscularly if the vertigo is severe. Since the cause of the hydrops is not known, various measures have been advocated to prevent the attacks. Restriction of salt and fluid intake is recommended. Vasodilator drugs have also been advocated. Vestibular sedative like cinnarizine 15 mgm three times a day is helpful if the patient can tolerate. If these measures do not help and the vertigo recurs often various surgical procedures have been recommended

such as section of the vestibular nerve or labyrinthectomy. The causes of vertigo are listed in Table 1.3.

TABLE 1.3
CAUSES OF VERTIGO

1. Benign positional vertigo
2. Vestibular neuronitis
3. Meniere's disease
4. Cerebello-pontine angle lesions (acoustic neurinoma)
5. Head injury
6. Cerebro-vascular disease and brain stem ischaemic attacks
7. Hyperventilation syndrome
8. Drugs-aminoglycosides, streptomycin, salicylates etc.
9. Psychiatric disorders.

Motor and sensory disturbances are described by the patients in a variety of terms. For example "heavy" and "numb" mean both weakness and loss of sensation. A weak limb is felt to be heavy as it is difficult to lift it against gravity. Loss of sensibility including posterior column sensation is often described as a limb being swollen and heavy. Limb weakness should be further localized into proximal, distal or global. **Proximal weakness causes the patient to complain of difficulty in lifting loads, combing hair and on going up the stairs.** Patients with distal weakness complain that objects are inadvertently dropped from the hand or that the great toe often gets stubbed. Muscle weakness resulting from nerve root entrapment is accompanied by pain and paraesthesias. Note whether or not there is atrophy. Are there any fasciculations? Is the muscle weakness symmetrical as in polyneuritis or asymmetrical as in poliomyelitis? Did the weakness come on suddenly, rapidly or gradually, and is it progressive? The following terms are generally used to describe motor dysfunction. Weakness of the limb is described as "paresis" whereas total paralysis of the limb is called "plegia". *Monoplegia* describes paralysis of one limb be it upper or lower limb, *hemiplegia* describes paralysis of one half of the body, *paraplegia* denotes paralysis of both legs whilst *quadriplegia* means paralysis of all four limbs. The term *diplegia* is used to describe paralysis of the legs that is secondary to cerebral pathology specially in infants.

Bilateral lesions of the corticospinal tract above the medulla cause *pseudobulbar palsy* (Bulb = medulla). It results in emotional incontinence

with pathologic laughter and crying, hoarse, explosive speech, dysphagia, a spastic tongue and bilateral pyramidal tract signs such as hyperreflexia and spasticity. Sensory complaints are described as a feeling of pins and needles - *Paraesthesia*, decreased sensation - *hypoaesthesia* or as an unpleasant sensation as burning pain—*dysaesthesia*. When sensory disturbances are due to peripheral neuropathy the sensory loss is distal in the areas of the hand and feet—the “glove and stocking” distribution. In radicular lesions the pain and decreased sensation occurs in the area supplied by the affected nerve root. Paraesthesias involving one limb or one half of the body are secondary to spinothalamic tract dysfunction.

We have already described the pain of headaches and its related syndromes. Neuralgic or neuritic pain is severe, shooting agonizing pain that occurs in bouts in sciatica due to prolapsed intervertebral disc and is worse when the patient is relaxed. A painful retro-orbital condition in which the eyes swell and there may be associated extraocular muscle palsies is the Tolosa-Hunt-Syndrome of painful ophthalmoplegia. It is more common in females and responds well to steroids. Causalgic pain is severe agonizing burning pain following nerve injury, most commonly after injuries to the sciatic or median nerves.

A very important commonly encountered symptom is transient loss of consciousness. It occurs in *seizures* and *syncope*. The word syncope is derived from the greek term synkope meaning to cut or break. In syncope there is sudden cessation of brain function due to a transitory reflex cerebral anoxia. The usual ‘faint’ also known as a ‘vasovagal’ attack can occur with very little provocation. Minor trauma, seeing unpleasant things, prolonged standing in one position, each of these can lead to syncope. There is a sudden fall in blood pressure with loss of vasomotor tone. Consciousness is regained rapidly if the patient is allowed to lie flat. Syncopal attacks are preceded by a feeling of lightheadness and nausea, the patient slumping to the floor from an upright position. The face is pale, the body bathed in sweat is cold and clammy. Rarely “convulsive syncope” occurs where there is brief limb jerking and sphincter incontinence. The causes of syncope are listed in Table 1.4.

TABLE 1.4
CAUSES OF SYNCOPE

- I. Postural hypotension
- II. Valsalva Manoeuvre (cough, micturition)
- III. Cardiac arrhythmias
- IV. Aortic stenosis
- V. Hypersensitive Carotid Sinus

In Stokes-Adams attacks the patient may fall heavily to the ground. *Postural hypotension* is defined as a fall in blood pressure which occurs in assuming the erect position. The symptoms are relieved by lying down. No agreement exists as to the normal range of postural changes in blood pressure but a consistent and persistent decrease of 25 mm Hg or more in diastolic and 15 mm Hg or more in systolic substantiates a clinical diagnosis of postural or orthostatic hypotension.

In generalized tonic clonic (grand mal) seizures there is loss of consciousness followed by jerking of the limbs; the eyeballs roll up, there is frothing at the mouth, the patient may bite his tongue and be incontinent of urine or faeces. The seizure lasts about two minutes and is followed generally by a postictal period of confusion or sleepiness which lasts up to at least half hour. The duration of the spell, the sequence of events, and the postictal phase help differentiate the seizure from syncope though at times the difference is not clear. In such a case additional history and special diagnostic tests such as the Holter monitor and Electroencephalogram (EEG) are needed. Gowers appropriately called this problem the "borderland of epilepsy".

INVOLUNTARY MOVEMENTS

Involuntary movements involving the face, neck, trunk or limbs are sometimes encountered. The commonest facial involuntary movements are associated with *chorea* where facial grimacing is observed. The patient appears to be making faces and when asked to protrude his tongue he is unable to keep it steady, the tongue keeps wriggling and popping in and out of the mouth. A complication of chronic psychotropic drug therapy is *tardive dyskinesia*. Coarse facial movements occur, the patient making sounds that resemble chuckling or sucking. Tardive dyskinesia is difficult to treat and is thought to occur as a result of sensi-

tization of dopamine receptors. The limbs and trunk may also be involved and dystonic posturing can be seen. *Facial myokymia* is characterized by involuntary unilateral fine continuous undulating movements of facial muscles. It can occur at any age, persist for months or years and has been seen in multiple sclerosis and tumors of the brain stem. *Facial myoclonus* may occur as a complication of Bell's palsy or may be idiopathic. Beginning as a flickering movement of the lower eyelid the movement spreads to involve the entire face. Though benign the movements are very distressing to the patient. It can be treated by tranquilizers in the initial stages. Partial neurectomy of the branches of the facial nerve has been found to be helpful in relieving the myoclonus. *Facial tics* consist of bilateral repetitive movements like blinking. They usually begin either automatically or as a result of some extrinsic inciting factor or an intrinsic stimulus like conjunctivitis. They are then perpetuated as a compulsion after the original stimulus has long ceased to operate. *Titubation* is an involuntary movement of the neck muscles and is generally a symptom of a systemic degenerative disorder. The movement occurs in the form of head nodding at a rate of four to five per second. *Tremors* are rhythmic regular oscillating movements of limbs, trunk, head or part thereof. *Fine tremors* of the fingers are seen in thyrotoxicosis or anxiety neurosis. Fine irregular tremors of the hands are also seen in lesions of the anterior horn cells or its axons. The *familial essential tremor* is coarser and may involve not only the fingers but the forearms producing rhythmic pronation-supination movements. The initial *tremor of parkinsonism* is described as a pill-rolling tremor. It is characterized by flexion and extension of the fingers with abduction and adduction of thumb. It occurs at the rate of four to eight cycles per second and is present only when awake. Aggravated when the patients attention is drawn to the limb and increased when the patient is emotionally disturbed, the tremor may come in the way of patients activities such as eating, and buttoning his clothes. In other diseases of the basal ganglia such as Wilson's disease the tremor may involve the proximal parts of the limb. This tremor is of large amplitude with regular movements of the upper limb, best seen when the shoulder is abducted and the elbows flexed. It resembles the wing flapping movements of a bird and is hence described as 'wing flapping tremor'. '*Rubral tremor*' is a rotatory type of tremor of the head or hands accompanied by random movements of the fingers and often abnormal head posture. It is often present at rest and made worse by movement.

Asterixis is seen in renal failure, hepatic failure and is not a true tremor. There is no alternate contractions of the agonist and antagonist

muscles, but there is alternate loss and regaining of tone in these muscles. Ballism is said to occur when the involuntary movements of the limb are unilateral, proximal, violent and flinging in character. They are described as *Hemiballismus* when they involve one half of the body. Some consider this to be a variety of chorea. The usual location of a lesion that causes hemiballismus is in or near the subthalamic nucleus (*Corpus Luysii*). Often the movements are so severe that if untreated the patient is exhausted and may have cardiac failure. These movements may respond to haloperidol, or reserpine. Stereotaxic surgery is indicated if the movements are not controlled by these drugs.

Chorea is a state of excessive, spontaneous movements, irregularly timed, non-repetitive, randomly distributed and abrupt in character. It may vary in severity from restlessness with mild intermittent exaggeration of gesture and expression, fidgety movements of the hands, and an unstable dancelike gait to a continuous flow of disabling violent movements. These movements hamper the activity of daily living. In the initial stages the patient's relatives are unable to recognize the true nature of the condition and feel that the patient is mimicking or imitating someone. In very mild cases the movements can only be observed when the patient is asked to stretch his hands. Hypotonia may accompany this movement disorder. Chorea may be unilateral or bilateral. *Sydenham's chorea* occurs in the second decade of life and may follow rheumatic fever. Rheumatic carditis may be associated with the chorea. Recurrences occur in about twenty per cent of cases. Chorea that occurs during pregnancy is known as chorea gravidarum. It is generally unilateral. Chorea is also known to occur in polycythaemia-vera, in systemic lupus erythematosus, and in certain heredo-familial degenerative disorders such as Huntington's chorea. Recently chorea has been described in women who have been using oral contraceptive pills. Haloperidol or Triperidol helps to control the choreic movements.

Athetosis is characterized by irregular, slow, forceful, writhing movements generally of the extremities, very often characterized by finger movements. The posture in athetosis is metaphorically described as a "policeman's hand". Kernicterus, anoxic disorders such as status marmoratus, and toxic disorders such as carbon dioxide and carbon disulfide poisoning can cause athetosis.

Dystonia Dystonic states are abnormal postures produced by slow sustained muscle contractions which distort limbs, trunk, neck, face or

mouth into characteristic postures; they may be distal (athetoid), proximal (torsion spasm) generalized, unilateral or focal. Typical dystonic postures include a hyperpronated forearm with a flexed wrist and extended fingers. The foot is inverted and plantar-flexed, the trunk curved to one side (scoliosis), backwards (lordosis), the neck rotated to one side (torticollis), forward (antecollis), or backwards (retrocollis). Dystonic posturing is seen in hereditary torsion dystonia and in association with other hereditary diseases such as Wilson's disease. It can also be seen secondary to head trauma or be drug induced. Stereotaxic surgery helps relieve dystonia.

Myoclonus is characterized by sudden shock-like contractions of a muscle or group of muscles. It may or may not be accompanied by joint movements and may occur at rest or during action. It may be isolated and consist of repetitive rhythmic movements as in palatal, or diaphragmatic myoclonus. A patient complains that he tends to drop things. It affects people physiologically at the onset of sleep, "sleep starts". It can be one of the manifestations of epilepsy and is seen also in severe progressive brain disease like subacute sclerosing panencephalitis, and in storage disorders such as Tay-Sach's Disease or in the post-anoxic encephalopathy of Lance-Adams that follows cardiac arrest.

Tics are sudden, stereotyped, complex, repetitive, normally coordinated but inappropriate movements, which can only be suppressed for a short period by an effort of will at the expense of increasing emotional tension. Tics may be single such as sniffing, vocalization, finger movements, or complex such as combinations of sniffing, and grunting and generalized where complex tics are accompanied by impulsive, emotional behaviour. Tics are seen in Gilles de la Tourette disease and sometimes following encephalitis.

PARKINSON'S DISEASE In 1917, James Parkinson wrote the classical clinical description of 'Shaking palsy'. In India, the disease begins before or around 50 years of age and has a male predilection. No specific etiological factor is known for the causation of this degenerative disease. Though, psychological trauma, head injury, infection etc. have all been blamed at one time or other. Nor is there as yet, any evidence to state that the disease is inherited. Clinical features like that of Parkinson's disease can also be seen in arteriosclerotic disease of the brain, following encephalitis, administration of reserpine or anti-psychotic drugs, and carbon monoxide or manganese poisoning.

The pathology consists of degenerative changes in the pigmented cells of substantia nigra and the locus coeruleus. Round hyaline bodies with a clear peripheral halo called Lewy bodies are seen in the cytoplasm of the pigmented nerve cells. There is also degeneration of nerve cells in the pallidum with loss or thinning of myelin sheaths of fibres in the ansa lenticularis, and striopallidal and cortico-pallidal fibres. In late cases, minimal cortical atrophy has been reported. Biochemically, secretion of dopamine, a neurotransmitter, is diminished in the striatum.

The disease may begin with the classical tremors described earlier in the chapter or rigidity. The tremors may start in one hand and progress to involve the lower limb on the same side and the limbs on the opposite side within a few months to few years. Rigidity initially involves the proximal muscles causing muscle and joint pains. Rigidity is of lead pipe or cog-wheel type affecting both flexors and extensors equally. It spreads slowly to involve all four limbs. The tremor may exist independent of rigidity, however, minimal slowness of movements (hypokinesia) will be demonstrable, and the patient may complain of altered handwriting. Finally, in a small number of patients, the disease may begin with dragging of one foot and lack of swinging of an arm.

The characteristic clinical features are the changes in facial expression and posture, tremor, rigidity, hypokinesia and altered gait.

The facial expression of a parkinsonian patient is characteristically immobile, fixed with absence of blinking and a stare, giving an impression of a mask (mask like faces). Smile is slow to develop. The blink elicited by rhythmic tapping on the bridge of the nose continues indefinitely in a parkinsonian (Gubellar tap sign). The speech is usually low pitched and monotonous and the sentence may end in a mumble. Dysarthria gradually worsens and the speech may be neither audible nor intelligible.

The patient is slow (Bradykinesia) and has loss of dexterity in performing movements. He also has difficulty in initiating movements (Akinesia) which at times, on meeting an obstacle while walking, makes him glued to the ground. It also leads to difficulty in writing, the handwritings becoming small (micrographia), and also produces difficulty in performing activities of daily living, such as buttoning, eating, tying shoe laces. The characteristic posture of parkinsonism develops later in

the course of the disease. The patient stands with flexion of the trunk and knees. The neck is held rigidly, and the arms are adducted, bent at elbow, with flexion and adduction of all fingers.

Initially, the patient walks with slow shuffling steps. Due to postural dysequilibrium and impairment of righting reflexes, patient may have anteropulsion (Festinant gait) or retropulsion, the former being common. In anteropulsion, he walks with quick shuffling steps giving an impression that he is about to fall forwards.

Autonomic disturbances such as excessive salivation, flushing of skin with sweating and constipation are usually present. Oculogyric crisis usually is seen in post-encephalitic parkinsonism. It consists of attacks of involuntary upward deviation of eyes with retraction of the upper lids.

The disease is slowly progressive without remission or exacerbation. Eventually the patient is bed-ridden though may live a normal span of life.

After introduction of synthetic drugs, Belladonna alkaloids are seldom used. Benzhexol hydrochloride (Pacitane) is given in the dose of 1 to 2 mgm. three times a day, and can be gradually increased to 12-15 mgm a day. Two other drugs available in India are Orphenandrine hydrochloride (Disipal) and Procyclidine hydrochloride (Kemadrine). The former is given in the dose of 50-150 mgm a day and the latter in the dose of 20-30 mgm a day, in divided doses.

After the introduction of levodopa (L-dopa), the treatment of parkinsonism is totally revolutionized. It has reduced the indications for surgery. The drug should be used with caution in patients with occlusive cerebral or coronary vascular disease, cardiac arrhythmias, renal and hepatic diseases, psychosis, haemolytic anaemia and glucose-6-phosphate dehydrogenase deficiency. The drug is marketed in India in 500 mgm scored tablets. Initially, 125 mgm two or three times a day is started adding 125 mgm once a week till desired response is obtained. The Indian population does not tolerate more than 3 to 4 G. of the drug per day. To avoid irritation of the stomach, the drug should always be given with or immediately after a meal. Postural hypotension and drug induced involuntary movements may restrict the use of the drug and the dose may have to be stepped down. Addition of decarboxylase inhibitor

(not available in India) prevents **utilization** of the drug by tissues other than brain, thus helping to reduce the total intake of levodopa.

Stereotaxic lesions are placed in the **ventro-lateral nucleus of the thalamus** and the subthalamic field of Forrel, to mainly reduce the tremors and rigidity. In patients, where tremors are not controlled by drug therapy and where the drug has produced involuntary movements consider surgery.

CHAPTER—II

EXAMINATION - HIGHER FUNCTIONS AND CRANIAL NERVES.

The higher functions of the central nervous system are examined in terms of the level of consciousness, orientation in time, space and person, memory, speech, judgement and insight. The level of consciousness can vary from the fully *conscious* state defined as a state in which there is awareness of the self and the environment, to the **comatose** state of unarousable unresponsiveness. States of altered consciousness include the **confusional** state where memory is faulty, but the person is oriented in time and space. **Delirium** is characterized by disorientation, agitation and visual hallucinations. The patient is fearful, talks loudly, and appears irritable and suspicious. **Stupor** implies a state of unresponsiveness in which the patient can be aroused with great difficulty. Terms such as obtundation, deeply stuporose, light coma, deep coma, make an accurate assessment of the level of consciousness difficult. In order to standardize the index of neurologic function the Glasgow Coma Scale has been introduced. This scale relates "consciousness" to motor responses, verbal responses and eye opening. Coma is defined as no response to command, no comprehensible verbal response, and no eye opening. In each of these categories the examiner determines the BEST response the patient can make to a set of standardized stimuli. The patient's response to the examiners stimuli is assigned a number as outlined below. The highest total score is fourteen the lowest is three. The Glasgow Coma Scale has proved to be very helpful in assessing and in monitoring the level of dysfunction especially in patients with head injury.

After ascertaining the Glasgow Coma Scale examine the fundus. Subhyaloid or preretinal haemorrhages may be seen in subarachnoid haemorrhage. The pupillary light reflex should be checked with a bright source. The reflex is preserved until the terminal stages in metabolic coma except when the coma is secondary to atropine or opiate overdose. When the pupils are unequal and non-reacting to light uncal herniation may have occurred. Pupils that are mid-positioned and fixed suggest a mid-brain lesion. In pontine lesions the pupils are very small but if examined under a magnifying glass the light reflex is usually

TABLE 2.1
GLASGOW COMA SCALE

Examiner's Test	Patient's Response	Assigned Score
Eye Opening		
a) Spontaneous	Opens eyes on own	4
b) Speech	Opens eyes when asked to in a loud voice	3
c) Pain	Opens eyes when pinched	2
d) Pain	Does not open eyes	1
BEST Motor Response		
a) Commands	Follows simple commands	5
b) Pain	Pulls a part of body away when examiner pinches patient	4
c) Pain	Flexes body to pain	3
d) Pain	Body becomes rigid in an extended position when examiner pinches patient	2
e) Pain	Has no motor response to pinch	1
BEST Verbal Response		
a) Speech	Carries on a conversation correctly and tells examiner where he is, who he is, and the month and year	5
b) Speech	Seems confused or disoriented	4
c) Speech	Intelligible articulation but speech is used only in an exclamatory or random way; no sustained conversation possible	3
d) Speech	Makes sounds that examiner cannot understand	2
e) Speech	Makes no sound	1

The highest level of response to each command is recorded by circling the appropriate number and the three categories are totaled.

present. Next examine the tone of the eyelids. Lift the eyelid and then release them. If the eyelid fails to close on either side it indicates involvement of the facial nerve on that side. Now test for blinking, either in response to light or threat. If blinking is absent on both sides it indicates pontine involvement. Check the corneal reflex with a stronger stimulus than would be used in an alert patient. Absence of the corneal reflex bilaterally is a grave sign implying absence of pontine function. Observe the position of the eyes at rest. Are there any spontaneous movement? Are the eyes conjugate or disconjugate? Roving eye movements are seen in comatose patients with an intact brain stem. The eyes move slowly from side to side. When spontaneous eye movements are absent the doll's eye manoeuvre should be performed. Stand at the head end of the bed and briskly rotate the head from side to side. If the eyes move in a direction opposite to the head the doll's eyes response is present. If no eye movement can be elicited by this manoeuvre then caloric stimulation should be done. First check the auditory canal to make sure that the tympanic membrane is intact. Now elevate the bed thirty degrees above the horizontal so that the lateral semi-circular canal is now vertical. Irrigate the eardrum with ice cold water. In a comatose patient the eyes will conjugately deviate towards the irrigated side and there may be a few irregular beats of nystagmus in the ipsilateral eye. Absence of eye movements even after caloric stimulation is a sign of severe brain stem dysfunction and is one of the signs of "brain death". Conjugate deviation of the eyes to one side or the other helps to localize the lesion. When there is a *destructive* lesion in one cerebral hemisphere the eyes "look" at the lesion that is they are deviated to the side of the lesion, the hemiplegia being on the opposite side. **Irritative** lesions cause the eye to deviate away from the affected hemisphere. In pontine lesions the eyes are deviated to the same side as the hemiplegia. When there is conjugate deviation of the eyes in the downward direction it implies compression in the region of the dorsal brain stem. It is seen in thalamic haemorrhage. Motor function is tested in terms of tone and power. Raise each limb individually against gravity and let it fall. Sudden uninhibited flopping of the limb indicates paralysis of that limb. Bend the leg at the knee and observe how the leg extends. If the leg extends gradually then there is no paralysis in that limb, if however, the leg flops right away it implies paralysis on that side. Generalized hypotonia or flaccidity occurs with metabolic coma. Observe the pattern of respiration. The breathing waxes and wanes. There is a gradual build up and then a period of apnoea. Central neurogenic hyperventilation occurs in mid-brain lesions. Apneustic breathing occurs with pontine lesion and implies a prolonged period of apnoea at the height of full inspiration.

Ataxic breathing or medullary breathing is irregular, ineffectual attempts at breathing. The causes of coma are legion and can be divided into neurological causes and metabolic causes. The table below enumerates some of the common causes of coma.

TABLE 2.2

NEUROLOGICAL CAUSES OF COMA

I Supratentorial lesions

- i. Infarctions
- ii. Haemorrhage
- iii. Neoplasms: Primary
Secondary
- iv. Abscesses, Granulomas.
- v. Trauma: Extradural haematoma
Subdural haematoma
- vi. Infections: Encephalitis, Meningoencephalitis.

II Infratentorial lesions

- i. Brainstem infarct
- ii. Brainstem haemorrhage
- iii. Brainstem tumour
- iv. Cerebellar haemorrhage
- v. Cerebellar abscess.
- vi. Infections—Meningitis.

METABOLIC CAUSES OF COMA

I Coma from diseases of the Brain

- | | | |
|------|-------------------------|--|
| i. | Grey matter diseases : | Presenile and senile dementias
Progressive myoclonic epilepsy
Lipid storage disease. |
| ii. | White matter diseases : | Schilder's disease
Leukodystrophies |
| iii. | Hypoxia : | Pulmonary disease
Decreased oxygen tension
Anaemia
Methemoglobinemia |

- | | | |
|-----|------------------------|--|
| iv. | Ischaemia : | Cardiac arrest, Stokes-
Adams syndrome, Myocardial
infarction and fallure
Aórtic stenosis
Syncope
Carotid sinus hypersensitivity
Low blood volume.
Hypertensive encephalopathy |
| | | Hyperventilation syndrome
Polycythemia |
| v. | Hypoglycemia | |
| vi. | Vitamin deficiencies : | Wernicke's encephalopathy,
Pellagra |

II Diseases of other organs

- | | | |
|------|--------------------------|---|
| i. | Hepatic coma | |
| ii. | Uremic coma | |
| iii. | CO ₂ narcosis | |
| iv. | Endocrinal diseases : | Symond's disease
Myxoedema
Addison's disease
Cushing's disease
Pheochromocytoma
Hyper and Hypo-parathy-
roidism |
| v. | Porphyrria | |

Cerebral infarctions rarely present as coma unless there is associated cardiac or pulmonary complications or electrolyte imbalance is present. The coma in cerebral infarctions is generally due to the occurrence of severe progressive cerebral oedema. The first indication that cerebral oedema is developing is a deterioration in the level of consciousness. The patient becomes dull and apathetic and soon is drowsy. A few become agitated. When coma ensues, the respiration becomes deep with yawning and intermittent pauses. The pupils are small, react to light and the doll's eye manoeuvre is positive.

A majority of intra-cerebral haemorrhages present with loss of consciousness. The haemorrhage may be secondary to a rupture of an

arteriosclerotic vessel or it can be secondary to bleeding from an aneurysm or angioma. A majority of the patients with cerebral haemorrhage have hypertension, the bleeding generally occurring when the patient is awake. The haemorrhage begins, the patient complains of severe headache and rapidly lapses into coma. In the initial stages a third nerve palsy may be seen because of uncal herniation. Later there is laboured respiration, a slow pulse and rising blood pressure. Finally the pupils dilate, are mid-positioned and fixed. The respiration becomes irregular, the blood pressure starts to fall, and is soon unrecordable. If there has been extensive haemorrhage involving the ventricle there is a sudden profound coma, death occurring within minutes or hours of the bleed because of ponto-medullary failure. A slowly progressive focal neurologic deficit should arouse suspicion of the space occupying lesion and modern investigative techniques should be employed to recognize the nature of the lesion. However, in certain instances coma can occur rapidly in a patient with a supra-tentorial tumour. Neoplasms tend to bleed, glioblastoma multi forme, haemangio-blastoma, and oligodendroglioma are tumours that are known to have sudden haemorrhages into their substance. Coma can also occur if a tumour is displaced suddenly obstructing the C.S.F. pathway as in the case of colloid cyst of the third ventricle. Metastatic lesions in the brain, when multiple can cause severe edema leading to early coma. Pontine haemorrhage occurs in hypertensives with the bleed happening during waking hours. The patient exhibits apneustic breathing, the pupils are pinpoint. The doll's eye manoeuvre may be absent. Flaccid quadriplegia with an extensor plantar response bilaterally may be seen. Examination of C.S.F. reveals blood. When the hypothalamic centers for thermal regulations are involved there are disturbances involving temperature control and hyperthermia may develop a few hours after such a haemorrhage. In coma secondary to brain stem infarction generally there is an antecedent history of transient ischaemic attacks. The patient must have previously complained of double vision, vertigo, difficulty with speech or numbness or heaviness of one or more limb. When coma ensues respiratory abnormalities occur which include periodic breathing, spells of hyperventilation, or even gasping for breath. The pupils may be either small or large and may not react to light. Hemiplegia or quadriplegia may occur and extensor plantar responses may be present. The patient may show *decerebrate* posturing on painful stimuli. When fully developed, decerebrate posturing consists of opisthotonus, the arms are extended and hyperpronated, the legs are extended, and the feet plantar flexed. Brain stem tumours rarely cause coma. Patients with

cerebellar haemorrhage develop coma suddenly. Some patients present with occipital headaches, vomiting, vertigo, and with deviation of the eyes to one side. Extradural haematoma in the supra-tentorial compartment is usually due to rupture of the middle meningeal artery. The middle meningeal artery is a branch of the external carotid. Many textbooks describe that this haematoma is generally associated with so called 'lucid interval' which actually is more often absent than present. Pupillary abnormalities precede the coma by a few hours. Initially the ipsilateral pupil is small because of parasympathetic irritation. It then dilates because of parasympathetic paralysis and sympathetic overactivity. Finally it becomes mid-positioned and fixed. This triad of pupillary changes is known as Hutchinson's triad. Sometimes the pupil dilates on the same side as the hemiplegia. This is known as the Kernohan's notch phenomenon. It occurs because there has been contra-lateral damage as a result of the cerebral peduncle being injured along the free edge of the tentorium. Few patients with a chronic subdural haematoma may present with coma. They generally have a history of headaches and confusion, and problems with memory prior to the development of the coma. One of the classical phenomenon of a chronic subdural is day to day and hour to hour fluctuations of the clinical picture. Grey and white matter diseases rarely progress rapidly and never cause a problem in the diagnosis of the coma. With cardiac arrest and carbon monoxide poisoning consciousness is lost if the cerebral circulation stops for longer than six minutes. Generalized convulsions may follow. The pupils are dilated, the plantars show bilateral extensor responses. If within this short period of time circulation is restored and consciousness returns then there is no resulting neurologic deficit. If the cardiac arrest lasts for more than four to six minutes the patient is left with substantial neurologic residue.

Severe anaemia, myocardial infarction, congestive heart failure, and pulmonary diseases lead to subacute or chronic hypoxia. The encephalopathy presents with confusion, disorientation, and drowsiness. Progressive hypoxia leads to periodic breathing, small non-reacting pupils, myoclonus, decorticate rigidity and hemiplegia. Hypertensive encephalopathy occurs due to sudden rise of blood pressure in a known hypertensive. It presents with focal or generalized seizures, headaches, vomiting and rapid progression into coma. Examination of the fundus reveals papilloedema with exudates. Monoplegia or hemiplegia with uni or bilateral extensor plantar responses may also be present. Hypoglycemia occurs if the blood sugar drops below forty milligrams and can present with seizures. Focal neurological signs can occur. There is initial

confusion and sometimes even a state of manic excitement. It is difficult to differentiate hypoglycemic coma from other causes of metabolic coma, hence, blood for sugar estimation should always be drawn in a case of coma prior to the administration of intravenous glucose.

Wernicke's encephalopathy occurs oftener than suspected. A patient with a past history of alcoholism who has a poor nutritional status, and problems with memory, who appears confused and disoriented on examination, should be carefully examined for nystagmus and oculomotor paralysis. If there is evidence of peripheral neuropathy then the diagnosis of Wernicke's encephalopathy is secure and intravenous thiamine should be promptly administered. Proper identification and treatment of this condition is very important since the patient can lapse into coma, have a fall in blood pressure and even die. Pellagra is seen often in our country because of nutritional deficiency. Skin changes and gastrointestinal disturbances along with peripheral neuropathy are seen in a patient who will present with delirium and confusion. Pyridoxin deficiency is usually seen in infants and children. They present with tremors and/or seizures and lapse into coma. Patients on Isoniazide therapy and hydralazine are also prone to develop Pyridoxine deficiency.

Hepatic Coma When a patient with acute or chronic liver disease becomes apathetic and quiet, or becomes delirious and manic, incipient hepatic coma should be suspected. Coma may supervene after a few days or may occur rapidly within hours. When the patient is obtunded, examination may show nystagmus, pupils that are small but react to light, and sometimes asterixis-involuntary movement of the hands, feet and, sometimes the tongue. They appear like flapping wandering movements. As the coma progresses the asterixis disappears. In the deeper stages of coma decorticate or decerebrate posturing may develop with rigidity and extensor plantar responses. Patients in hepatic coma have hyperventilation because of alkalosis. Focal neurological signs like hemiplegia are rare and convulsions are even rarer. If focal neurological signs are present the coma can be mistaken for a structural lesion of the brainstem. However, doll's eyes movements are present and the pupils show no abnormality. The serum ammonia is raised. The PCO_2 is low and the CSF is normal in hepatic coma. The electroencephalogram (EEG) may show the classical triphasic waves. These waves are transient and unless serial EEGs are done they may not be observed on a single EEG recording. In early coma the EEG will show bifrontal symmetrical slow activity and in deeper coma the background shows diffuse slowing in the delta range.

Uremic Coma In uremic Coma, tetany, myokemic irritability, myoclonus and seizures are common. Focal neurological signs like monoplegia or hemiplegia may be present. In the early stage of coma asterixis may be seen. The tendon jerks are hyperactive and the plantar responses extensor. Acidotic breathing similar to that seen in diabetic coma is present. Uremic coma may be difficult to distinguish from hypertensive encephalopathy. Blindness and papilloedema does not occur in uremic coma but does occur in hypertensive encephalopathy. The CSF pressure is raised in uremic coma while it is normal in hypertensive encephalopathy. **Diabetic Coma** Ketotic and non-ketotic diabetic acidosis generally develops in an uncontrolled or poorly treated diabetic. It may at times be the presenting symptom of diabetes. Generally there is a history of anorexia, nausea, vomiting and abdominal pain. The patient may have a mild elevation of temperature, the face is flushed, the skin dry. The respirations are long, deep and slow. The pupils may be large but react to light. No focal signs are present and the tendon reflexes may be depressed. It is important to differentiate diabetic coma from hypoglycemic coma since both types of disturbances can occur in the same patient. Vomiting is absent in hypoglycemic coma. The face is pale, the skin is moist and sweating may be excessive. There may be initial tremors and later focal neurologic signs may be present. The respirations in hypoglycemic coma are shallow and rapid. The table below presents the differentiating features of diabetic coma and hypoglycemic coma.

TABLE 2.3

	Hyperglycemic coma	Hypoglycemic coma
Onset	Slow	Rapid
Food	On food	Off food
Thirst	Marked	Absent
Hunger	Absent	Marked
Vomiting	Present	Absent
Skin	Dry	Moist
Face	Flushed	Pale
Tremors	Absent	Present
CNS signs	No focal signs	Focal signs present
Respiration	Deep, slow	Shallow, rapid
Acidosis	Present	Absent.

Barbiturates and other sedative drug poisoning During the drowsy state nystagmus, ataxia and dysarthria are present. As the coma deepens the doll's eyes manoeuvre fails to show eye movement, the pupils are small but react to light, and caloric testing shows depressed or absent response. There is generalised flaccidity, the deep tendon jerks are hypoactive or absent, respirations are slow, and the blood pressure falls.

EXAMINATION OF AN ALERT PATIENT

The assessment of an alert individual begins with tests of cognitive function. Many specific tests of cognition are usually unnecessary in routine neurologic examinations. If the patient is able to give a detailed account of his illness in a chronological order, it clearly indicates that he has normal memory and orientation. Mood and attention span can also be assessed during the interview. Gestures, grimaces, tics, and other involuntary movements may be observed. Whilst the patient narrates the history, closely observe whether there is any thought disorder. Delusions of grandeur, hallucinations of taste or smell may be present. The patient's insight and judgement can be assessed by his or her attitude towards the present illness, and the resultant financial and emotional strains that result from it.

Memory is further assessed by asking the patient to recall some recent events such as the date of admission to hospital. The general information and past memory can be tested by asking the patient to recall milestones in Indian history such as the year of India's independence. If the patient is illiterate ask him to recall past events in his own life, such as the ages of his children etc.

Tests for recent memory include the digit span test in which the patient repeats a series of numbers forwards and backwards. Begin by asking the patient to repeat 3-4 digits. A normal person should be able to repeat 7 digits forwards and 4 digits backwards. Simple calculations such as subtracting 7 from a 100 is then done. The patient is asked to subtract 7 from 100 and then to continue to subtract 7 from the result in a serial manner. Tests for cognitive function are specially important in dementia and parietal lobe lesions.

LANGUAGE Is defined as the art of communication of ideas and thoughts by means of symbols either written or spoken. Disorders of language due to lesions of the cerebral speech areas are known as *aphasias*. Disorder of spoken speech due to affection of the peripheral

mechanisms of speech production is known as **dysarthria**. If vocalisation is absent due to laryngeal involvement-aphonia-the patient retains the ability to comprehend and written speech is normal.

For clinical purposes it is convenient to divide aphasias into fluent and nonfluent depending on the characteristics of the language disorder. The aphasia is also dependant on the site of the lesion : anterior or posterior to the Rolandic fissure (see figure 2.1).

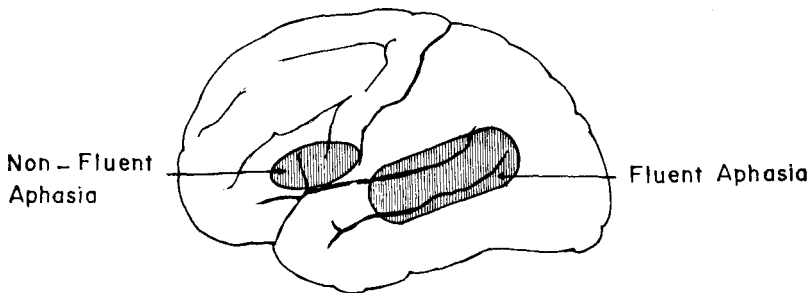


Fig. 2.1 : Lesions producing non-fluent & fluent aphasias

Aphasias are further classified depending on the patient's ability to repeat spoken speech (Fig 2.2). The table below classifies the clinical varieties of aphasia.

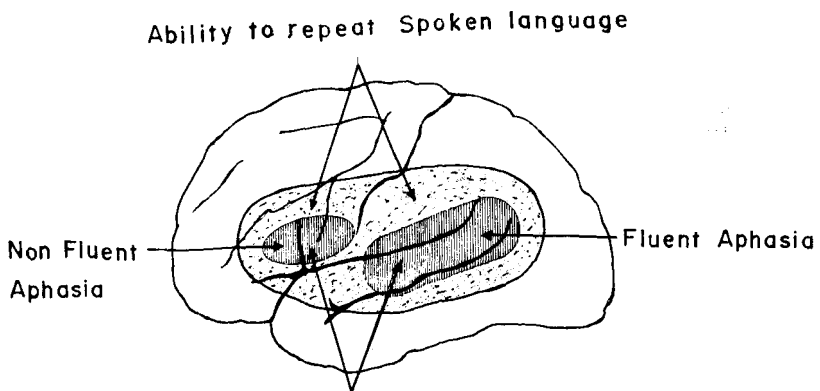


Fig. 2.2 : Inability to repeat spoken language.

TABLE - 4

CLINICAL VARIETIES OF APHASIA

A. Aphasia with repetition disturbance

1. Broca's aphasia
2. Wernicke's aphasia
3. Conduction aphasia

B. Aphasia without repetition disturbance

1. Isolation of the speech area
2. Transcortical motor aphasia
3. Transcortical sensory aphasia
4. Anomic aphasia

C. Disturbance primarily affecting reading and writing

1. Alexia with agraphia

D. Total aphasia

1. Global aphasia

E. Syndromes with disturbance of a single language modality

1. Alexia without agraphia
2. Aphemia
3. Pure word deafness

F. Non- aphasia misnaming

Broca's aphasia is a non-fluent aphasia, which manifests as stammering in its minor form, and as short, hesitant, telegraphic speech in its major form. Speech output is reduced to 5-10 words per minute. Even these words are enunciated with great effort, the patient resorting to facial grimacing, body movements, and gesturing to convey his thoughts or intentions. *Dysprosody* describes the unmelodic, arrhythmic, dysarthric speech of a non-fluent aphasic. In its most severe form the patient's vocabulary may consist of a single word which is used repeatedly, and incorrectly. The lesion is in the posterior frontal lobe. In lesions of Broca's area there is inability to repeat spoken speech. Comprehension of spoken and written speech is preserved.

In fluent aphasia, also known as sensory aphasia or Wernicke's aphasia the word output is higher than normal at 100 - 200 words per

minute. They come out effortlessly, with correct intonation and rhythm. However there may be pauses during speech with words inserted randomly producing no meaningful speech. Paraphasia—generation of words by substitution of syllables—and paragrammatism—faulty grammar—are present. Depending upon the extent of the lesion the patient may have only babbling or baby speech. Understanding of spoken and written language is also affected.

Other types of aphasia include conduction aphasia, pure word deafness, and pure word blindness. In Wernicke's aphasia the lesion is in the posterior superior temporal gyrus, in conduction aphasia the perisylvian region is affected, with the aphasia showing features of both fluent and non - fluent varieties in that there is difficulty in comprehension and repetition.

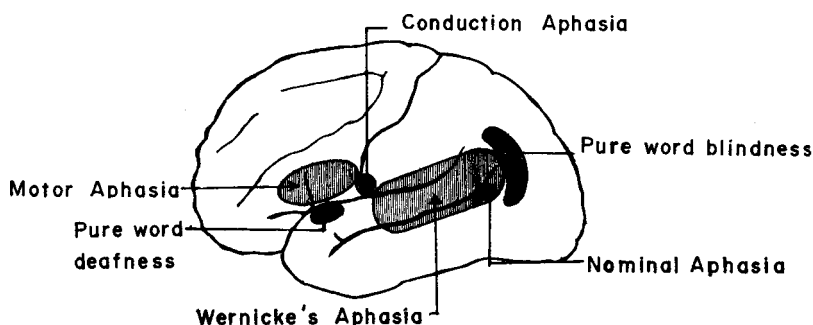


Fig. 2.3 : Schematic diagram showing lesions producing different types of aphasia

In pure word deafness the reception and understanding of spoken speech is affected with intact comprehension of written language. This form of aphasia is extremely rare, the lesion being in the dominant Heschl's gyrus. In pure word blindness on the other hand there is difficulty in comprehension of written language with preservation of the understanding of spoken speech. The lesion is in the parieto-occipital region. *Alexia* means an inability to understand and read written language. In its minor form it is known as dyslexia. In developmental dyslexia a child has difficulty reading at an early age. In alexia without agraphia there is inability to read, whilst the ability to write is unaffected. The lesion is in the dominant occipital lobe with involvement of the splenium of the corpus callosum.

In nominal aphasia the patient's speech appears normal, comprehension of spoken and written speech is normal, but there is difficulty in naming objects, for instance when shown a pen, the patient calls it a 'nab' or 'nen' but when offered the correct name he immediately accepts it. On being shown another object he persists with the word 'pen' though he is aware that he is wrong. This is known as perseveration. The lesion is said to be localisable in the dominant angular gyrus, though nominal aphasia is more often not localisable, being seen even in thalamic aphasia. In global or central aphasia the lesion is extensive, involving the frontal, parietal, and temporal speech areas on the dominant side. There is difficulty in understanding spoken and written speech as well as an absence of speech. The entire mechanism of language formation is deranged. Such massive lesions are generally seen with tumours.

An aphasic patient should first be observed carefully. Note the word output and rhythm. Ask the patient to repeat, read and write. Check to see if comprehension is impaired or not. Detailed aphasic testing requires patience and skill and may only be completed after several sessions.

CRANIAL NERVES

Olfactory Nerve (First Cranial Nerve) The filaments of the olfactory nerve arise from the mucous membrane of the nasopharynx and end in the olfactory bulb in the base of the anterior cranial fossa underneath the frontal lobes. Second relay fibres arise from the bulb and proceed caudally as the olfactory tract and end in the uncus and hippocampal gyrus, the cortical centre for smell.

To check the sense of smell the patient is asked to close his eyes and recognise different smells. One nostril is tested at one time, the other nostril being blocked by the examiner's finger. Perfumes, camphor, menthol, coffee etc are held near the open nostril and the patient is asked to inhale slowly and identify the substances. The sense of smell may be affected due to local lesions of the nasal mucosa as well as lesions of the olfactory nerve and its pathways. Head injury and olfactory groove meningioma are the commonest causes of loss of the sense of smell. Head injury may result in an altered sense of smell called parosmia where strong odours acquire an unpleasant quality. Hallucinations of smell arise in some type of epilepsy where the discharge arises in the uncinate region of the temporal lobe.

The Optic Nerve (Second Cranial nerve) The fibres of the optic nerve arise from the retina and exit at the optic nerve head where they acquire a myelin sheath. The optic nerve head or optic disc appears as a pinkish circular disc on fundoscopic examination. The nerve proceeds to enter the cranial cavity through the optic foramen of the sphenoid bone. Both optic nerves then proceed caudally and medially to meet and form the chiasm. Fibres from the nasal half of the retina decussate. The optic tract then arises and travels posteriorly to the lateral geniculate body. Fibres subserving the pupillary light reflex do not synapse in the lateral geniculate body and leave the visual pathway to travel towards the quadrigeminal plate of the midbrain. The visual fibres synapse in the lateral geniculate body and a second relay of fibres from optic radiation arise and travel in the posterior limb of the internal capsule to terminate in the visual cortex. The decussation of nasal fibres results in each occipital lobe controlling temporal fibres of the same side and nasal fibres of the opposite side. Lesions of the optic nerve lead to diminution of acuity of vision and to scotomas which are defined as islands of loss of vision. Lesions of the optic pathways posterior to the chiasm lead to defects in perception of certain parts of the visual field. When there is a defect in one half of the visual field it is called a hemianopia.

Optic neuritis and papilloedema lead to a decrease in the acuity of vision for distant objects. Loss of vision for near objects occurs in ophthalmic conditions. Visual acuity is tested by the Snellen's chart. Letters of decreasing size are arranged in rows on the chart with the largest letters on the top. The size of the letters is standardized so that the top row of letters can be read at a distance of 60 meters by a person with normal vision. The next row of letters can be read at 36 metres, the row below it at 12 metres and the last row at 6 metres. Since a person with normal vision can read the smallest letters at a distance of 6 metres his acuity is expressed as 6/6, whilst a person with a marked decrease in visual acuity may have an acuity of 6/60. If the person is unable to read the chart at all, acuity is then assessed by asking him to count fingers.

At the bedside the field of vision is tested by the "confrontation method". The examiner's vision is taken to be normal as compared to the patient. The examiner sits or stands in front of the patient at a distance of about 3 feet. The patient is asked to cover one eye and the examiner then covers his own eye, the eye that is directly opposite to the patient's covered eye. The patient is then asked to look at the examiner's eye. A small object held in the examiner's hand is slowly

brought into the field of vision from the periphery. The patient is asked to indicate the time he perceives the object. Each quadrant is tested individually. If the patient perceives the object at the same time as the examiner his field is said to be normal

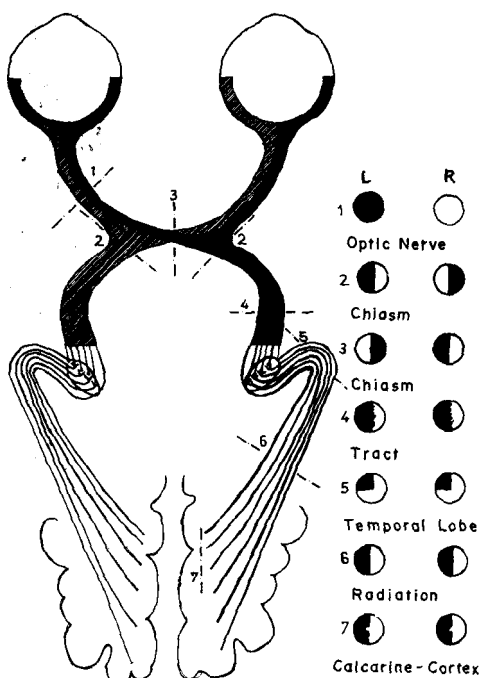


Fig. 2.4 : Schematic diagram of visual pathway and type of loss due to lesions at various levels

TABLE 2.5

1. Optic nerve	a. Retrobulbar neuritis b. Papilloedema c. Glioma and other tumours d. Optic neuritis due to various causes.	Loss of vision in one eye.
2. Chiasmal lesions	a. Tumours of Pituitary b. Hydrocephalus	Bitemporal Temporal field defects.

3. Chiasmal lesions	a. Tumours in and around chiasma b. Pituitary tumours c. Carotid artery aneurysm d. Arochnoiditis-Tuberculous.	Bilateral nasal field defects.
4. Tract lesions	a. Vascular accidents b. Tumours of temporal lobe c. Injuries	Incongruous hemianopia with macular sparing
5. Radiation lesion	a. Tumours of temporal lobe	Upper quadrantic field defect
6. " "	a. Cerebro vascular accidents. b. Abscess in the temporal lobe c. Head injury	Congruous hemianopia No macular sparing
7. Calcarine fissure lesion.	a. Cerebro vascular accidents b. Tumours	Congruous hemianopia with macular sparing

In children and obtunded patients the field of vision is tested by the 'menace test'. 'Reflex blinking' occurs when a moving object suddenly appears before eye. To test different quadrants the examiner brings his hand in suddenly from different directions and presence or absence of blinking is noted.

In parietal lobe lesions, the patient may be unable to perceive objects in one field -*visual inattention*- in the presence of a normal visual field. Visual inattention is assessed by asking the patient to keep both eyes open. The examiner stands opposite the patient and moves both hands simultaneously. The patient is then asked which hand is moving. The patient with visual inattention is unable to perceive movement in both fields and ignores one side. Rarely a lower quadrantic field defect may be present.

The different types of field defects that are commonly seen are listed in the Table 2.5. Loss of vision in one visual field is termed hemia-

nopia, which may be unilateral or bilateral. In bilateral loss involving the temporal half of one side and the nasal half of the opposite side, the deficit is known as a *homonymous heminopia*. This is further described as congruous or incongruous depending upon the visual field loss being symmetrical and equal or asymmetrical respectively on both sides. The extent of visual field loss differs for different colours being maximal for white and minimal for green.

Ophthalmoscopic examination An instrument called the ophthalmoscope is used, and the examination is an essential part of the neurological examination. It allows the examiner to view the optic disc or the nerve head, the blood vessels, the retina and the macula. The disc may reveal optic atrophy or papilloedema. In retro bulbar neuritis no changes are discernible on ophthalmoscopic examination. The blood vessels may show arteriosclerosis, arteritis, cholesterol emboli, and aneurysms in diabetes. Retinal examination may show retinitis pigmentosa, retinal detachment, or chorioretinitis. Subhyaloid haemorrhage is a preretinal haemorrhage seen in subarachnoid haemorrhage. A macular lesion known as the cherry red spot is seen in lipidosis. The macular flare and star are seen in hypertension and renal disease.

The causes of optic neuritis are listed in the Table 2.6

TABLE 2.6

CAUSES OF OPTIC NEURITIS

- | | |
|------------------------|--|
| 1. Inflammatory | Arachnoiditis
Meningitis-tubercle, syphilis
Tabes and tobo-paresis.
Inflammation of the orbit, eye and
cavernous sinus thrombosis. |
| 2. Toxic | Clioquinol
Tobacco
Methyl alcohol
Heavy metals
Carbon disulphide
Aniline dyes
Quinine. |

- | | |
|--|---|
| 3. Deficiency & Metabolic disorders | Vitamin B Complex group deficiency
Vitamin B 12 deficiency
Leigh's encephalomyelitis
Diabetes. |
| 4. Trauma | Head injuries. |
| 5. Vascular | Central retinal artery thrombosis
Cavernous sinus thrombosis.
Temporal arteritis.
Aneurysms. |
| 6. Compression | Craniostenosis
Paget's disease of bone (rare in India)
Optic nerve glioma
Meningioma of the olfactory groove.
Pituitary tumours.
Hydrocephalus
Post papilloedematous. |
| 7. Heredo-familial disorders | Leber's hereditary optic atrophy
Lipidosis
Retinitis Pigmentosa
Hereditary ataxias-Freidreich's |
| 8. Demyelination | Multiple sclerosis
Post infectious-following measles, chicken pox etc.
Schilder's, Devic's disease
Retrobulbar neuritis |

Papilloedema may occur due to increased intracranial pressure from tumours, blocks in the CSF pathway, hydrocephalus, and cerebral oedema. It is also seen in malignant hypertension, central retinal vein thrombosis, and in metabolic disorder such as hypercapnia, hyperparathyroidism and sometimes in the Guillain-Barre syndrome.

When the optic neuritis causes swelling of the nerve head, it may be difficult to differentiate from papilloedema. The following points help in the differentiation.

TABLE 2.7

	Optic neuritis	Papilloedema
1. Onset	Acute	Gradual
2. Vision	Total loss Central Scotoma Good recovery	Gradual diminution Peripheral constriction Partial recovery
3. Colour vision.	Red more affected	All colours equally affected.
4. Pain	Present	Absent
5. Vessels	Arteries narrow Veins congested.	Arteries normal Veins congested.

A recent advance in electrophysiology allows the estimation of the visual evoked response and detects abnormalities in the optic nerve. It has been of great help in the diagnosis of multiple sclerosis. The technique involves the recording of responses from the occipital region following stimulation of one eye with a flickering checker board pattern on an illuminated screen. The time taken for the pattern to be recognised by the occipital lobe is compared to standardized norms. A delay in recognition points to a defect along the optic pathway.

The Oculomotor (third), Trochlear (fourth), and Abducent (sixth) nerves The oculomotor nerve arises from an elongated nucleus lying rostrally to the aqueduct of Sylvius in the midbrain. Fibres for the levator arise from the caudal end of the nucleus while the ciliary fibres arise at the rostral end. The inferior rectus, inferior oblique, internal rectus, and superior rectus are represented in the nucleus in descending order. The fibres pass through the red nucleus and the substantia nigra to emerge at the interpeduncular fossa.

At the tentorium the third nerve lies between the posterior cerebral and superior cerebellar arteries. It then traverses anteriorly to lie in the lateral wall of the cavernous sinus. It enters the orbit through the superior orbital fissure after dividing into two branches. It supplies all the muscles of the eye except the superior oblique and the lateral rectus, and it also carries fibres for the constrictor muscles of the pupil.

The fourth or trochlear nerve arises from a nucleus situated below the third nerve nucleus, in the midbrain, ventral to the aqueduct of Sylvius. The fibres decussate at the level of the inferior colliculus. They course backwards to emerge dorsally, then course around the cerebral peduncle and come to lie in the lateral wall of the cavernous sinus. The nerve enters the orbit through the superior orbital fissure and supplies the superior oblique muscle.

The sixth or abducent nerve arises from a nucleus in the dorsolateral aspect of the pons near the floor of the fourth ventricle. Fibres of the seventh nerve wind around the nucleus of the sixth nerve, a matter of clinical relevance. Fibres of the abducent nerve then course downwards and forwards to emerge at the junction of the pons and the medulla. The nerve then has a long course at the base of the brain. It pierces the dura, travels below the petrosphenoidal ligament, and enters the cavernous sinus. It enters the orbit through the superior orbital fissure and supplies the lateral rectus muscle.

These three cranial nerves are generally examined together because they supply the extraocular muscles which work in pairs. The medial rectus of one eye works in tandem with the lateral rectus of the other eye. The two medial recti work together to produce convergence. The eye is moved upwards by the superior rectus and the inferior oblique, the former working optimally when the eye is adducted by the medial rectus. Similarly downward gaze is brought about by the actions of the inferior rectus and the superior oblique. The former functions well when the eye is abducted and the latter when eye is adducted. The movement of the eyes is under cortical control. Lateral gaze is under the control of centres located in the frontal and parieto-occipital regions. The frontal lobes control voluntary eye movements in horizontal gaze. These movements are rapid, and occur in either direction. These rapid eye movements are known as *saccades*. Saccadic eye movements occur in a "pulse-step" manner. *Pulse* is a burst of high frequency activity that precedes eye movement. Once an image is brought to focus at the fovea, it is maintained there by smooth slow movements known as *slow phases*. The 'step' holds the eye in the new position against orbital elastic restoring forces. The tracking of a moving object is called smooth pursuit. *Pursuit* is controlled by the parieto-occipital area in close conjunction with the visual cortex.

Voluntary conjugate gaze movement Fibres subserving conjugate

gaze arise from area 8 in the middle frontal gyrus. The fibres descend in the anterior limb of the internal capsule to the pons where they

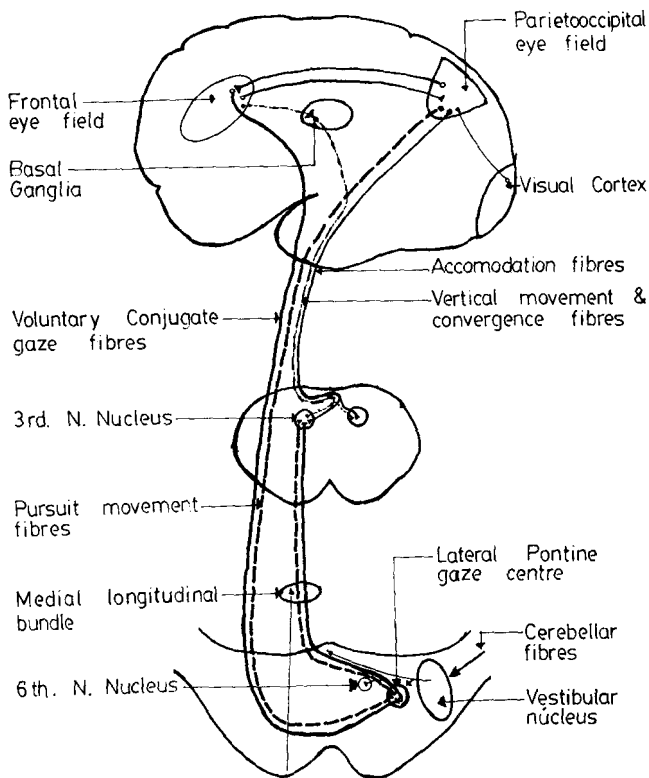


Fig. 2.5 : Centres and pathways of eye movements

decussate. They end in the centre for conjugate gaze now known by the name of P.P.R.F. or the para pontine reticular formation. The adjacent nucleus of the sixth nerve is also controlled by this centre. Fibres from the sixth nerve cross over to the opposite third nerve nucleus and travel in the medial longitudinal fasciculus. This allows both eyes to work in conjunction, the right frontal lobe initiating movement to the left and the frontal lobe to the right.

Vertical gaze requires input from both frontal lobes. Covergence is controlled by an area in the periaqueductal grey matter. Impairment of upward gaze occurs in Parinaud's syndrome which results from lesions of the pineal body and posterior third ventricle. The downgaze centre is not well localised but neurons in the pretectal region ventral to the aqueduct may control this movement. (Fig 2.5)

Pursuit eye movements In these movements the eyes move with the moving object and keep the object focussed on the macula or the fovea centralis. The fibres arise from the area lying anterior to area 19. They also end in the conjugate gaze centre travelling via the tectum and tegmentum of the midbrain and pons. Pursuit eye movements are located in the ipsilateral parieto-occipital cortex. Additional fibres from the frontal lobes, auditory and visual areas also play a role in mechanism of gaze. (Fig 2.5).

In a complete third nerve lesion, the affected eye is closed due to ptosis. When the lid is lifted the eye is deviated outwards and downwards due to the unopposed actions of the lateral rectus and superior oblique respectively. To test the integrity of the fourth and sixth nerves the examiner tests lateral gaze in the unaffected eye and observes whether diplopia disappears in that direction. The normal downward movement of the superior oblique cannot be tested as the paralysed medial rectus does not allow adduction of the eye. The secondary action of the superior oblique is observed when the patient tries to look downwards. The eye rotates inwards due to the pull of the superior oblique. This rotatory movement provides evidence of intact fourth nerve function. In a third nerve lesion the pupil is dilated and does not react to light. This is secondary to involvement of the parasympathetic fibres supplying the pupil.

An isolated lesion of the fourth nerve causes weakness of the superior oblique muscle only. When the patient looks straight ahead the

affected eye is rotated minimally outwards producing diplopia with the images on top of one another. To avoid the false image the patient tilts his head slightly away from the side of the affected eye. The diplopia is worse when the patient tries to look down as in reading or going down the stairs.

In an isolated lesion of the sixth nerve, the eye is drawn inwards. To avoid diplopia, the patient may involuntarily close the normal eye giving the impression of ptosis in the unaffected eye. When the affected eye is closed voluntarily, the 'ptosis' in the normal eye disappears. The patient avoids diplopia by turning the head in the direction of the affected eye.

Paralysis of the ocular muscles can occur due to a variety of causes. Congenital paralysis of one or more ocular muscles occurs due to absence of the muscle or defects in the nuclei. The lateral rectus is the most commonly absent muscle. 'Moebius Syndrome' results in bilateral abducent palsy with facial diplegia. Associated abnormalities include atrophy of the anterior part of the tongue, hearing loss, club foot and other skeletal abnormalities.

Head trauma may lead to third, fourth or sixth nerve paralysis. The sixth nerve is most commonly involved, followed by the third and fourth. Paralysis may occur due to a lesion in the brainstem, or lesions in the nerve anywhere along their course. Vascular lesions of the brainstem, multiple sclerosis, pontine gliomas, and Wernicke's encephalopathy lead to lesions that involve the nuclei of the oculomotor, trochlear and abducent nerves.

In our country, tuberculous meningitis is a common cause of ophthalmoplegia. Bacterial and fungal meningitis, meningo-vascular syphilis, sarcoid, and the Guillain-Barre syndrome are other causes of ophthalmoplegia secondary to nerve involvement. Nasopharyngeal carcinoma invading the base of the skull and pituitary tumours are some of the tumours that lead to paralysis of the extra ocular muscles. A common, and important cause of third nerve palsy is an aneurysm of the posterior communicating artery. All three nerves may be involved in a lesion of the cavernous sinus. Aneurysm of the internal carotid, and cavernous sinus thrombosis may cause a total ophthalmoplegia.

These nerves are also involved in lesions in or around the superior orbital fissure, and the orbit. Lesions causing muscle involvement may

TABLE 2.8

Site of lesion	Cause of lesion	Nerve involved
Nuclear	Brain-stem vascular disease	Any
	Pontine glioma	any or all
	Multiple sclerosis	any
	Poliomyelitis	any
	Wernicke's	Commonly third
	Congenital absence	Commonly third or sixth, 4th rare.
Posterior fossa	Meningitis (TBM, fungal bacterial or syphylitic or carcinomatous)	Commonly sixth or third.
	Nasopharyngeal Ca	Any
	Sarcoid	Any
	Guillain-Barre	All
	Herpes Zoster	Any
	Anuerysm of basilar or posterior communicator	Third
	Increased Intracranial tension	Sixth
	Migraine	Transient any or all
Lesions at petrous tip	Mastoiditis	Sixth
	Gradenigo's syndrome	Sixth with 7th, 8th and 5th
	Carcinoma naso-pharynx	All
Lesions in the cavernous sinus	Cavernous sinus thrombosis	Sixth later third
	Intrasellar tumours	Third
	Aneurysm carotid	Third or sixth.
Lesions at superior orbital fissure and orbit.	Tumours of orbit	Any or all
	Inflammatory lesions	Any or all
	Pseudo-tumour orbit	Any or all

simulate nerve and nuclear lesions. Of these, the most important ones are myasthenia gravis, progressive ocular myopathy and thyrotoxicosis. Fourth nerve lesions are very rare and occur in diabetes mellitus, head trauma, and in a common tumour of childhood, the medulloblastoma. The table 2.8 lists the causes of lesions involving the third, fourth and sixth nerves with regard to site.

The pupils The constrictor fibres of the pupils are derived from the parasympathetic system, while the dilator fibres are supplied by the sympathetic nervous system. The size of the pupils at rest depends on the intensity of light falling on either eye and on the parasympathetic nerves. The size, shape, and equality of the pupils have to be noted. The direct light reflex is elicited by using a bright light. Be sure you have checked that the flash light is functioning well. A poor source of light may fail to elicit the light reflex and lead to a false assessment of the patient. On shining a light in the eye, the pupil will constrict briskly. This is the direct light reflex. The indirect light reflex or the consensual light reflex is the simultaneous constriction of the opposite pupil. Accommodation is tested by asking the patient to look at his nose and observing the pupils constrict as the patient tries to converge the eyes.

If the pupils are unequal in size, decide which is the abnormal side. If there is ptosis on the side with the larger pupil the patient has a partial third nerve lesion on this side. If however, ptosis is present on the side of the smaller pupil, the patient may have Horner's syndrome. Both the light and accommodation reflexes are normal in Horner's syndrome.

The light reflex Impulses generated by light falling on the retina are conveyed by the optic nerve to the chiasm. Here nasal fibres cross and both optic tracts now carry the message to the lateral geniculate bodies. Fibres subserving the light reflex are then relayed to the Edinger-Westphal nuclei in the midbrain. Light falling on one eye causes bilateral excitation of these nuclei, hence there is both a direct and consensual response to light. Fibres from these nuclei travel with the third nerve to the orbit; lying in the superior aspect of the nerve. The pupillary fibres are located superficially. They finally relay to the ciliary ganglion and supply the constrictor muscle of the iris.

Accommodation reflex Nuclei that control the movements of the medial recti lie adjacent to the Edinger-Westphal nuclei. When the patient tries to converge he contracts both medial recti. Impulses from the nuclei of the medial recti overflow to the Edinger-Westphal nuclei causing the pupils to constrict on convergence.

Lesions of the light reflex pathways In cortical blindness due to bilateral cortical lesions, the pupils react normally to light. Fibres subserving the pupillary light reflex are unaffected as they separate from the visual pathways at the level of the lateral geniculate bodies. In bilateral optic nerve lesions, the pupils are dilated and fixed i.e. they do not react to light. In unilateral lesions of the optic nerve the pupillary size is unaffected because of the consensual light reflex. Lesions that compress the midbrain may result in pupils that are fixed and dilated. The Marcus-Gunn phenomenon or the swinging flash light test is useful in detecting lesions of the afferent arc of the light reflex. When light is shone on the normal eye both pupils constrict briskly. When the light is moved to the affected eye, the pupil paradoxically dilates. This is secondary to a lesion in the afferent pathway, generally the optic nerve. The pupil of the affected eye can constrict consensually because the efferent pathways are still intact. This pupillary abnormality is seen in retrobulbar neuritis.

Argyll-Robertson pupils are secondary to a lesion along the light reflex pathway at the level of the periaqueductal grey matter of the upper midbrain. The pupils are small, irregular, do not react to light, but react to accommodation. There is no pupillary dilatation on instillation of atropine.

Lesions of the third nerve that affect the pupil usually indicate a 'surgical' lesion such as an aneurysm, whilst lesions that spare the pupil are due to 'medical' causes such as diabetes. Pupillary fibres may be affected in diphtheritic paralysis. Associated palatal paralysis may be present.

Lesions of the sympathetic pathway The sympathetic system originates in nuclei situated in the hypothalamus. Fibres descend to the thoracic spinal cord where they synapse. Second order neurones now send their fibres to the superior cervical sympathetic ganglion. Third order neurones send their fibres with the carotid artery to enter the skull. They enter the orbit with the nasociliary branch of the fifth nerve to innervate pupils.

Sympathetic fibres also innervate Mueller's muscle, the smooth muscle of the eyelid. A lesion anywhere along this long pathway can result in a Horner's syndrome. The syndrome consists of ptosis, miosis, and anhidrosis. The eye on the affected side may appear to be shrunken (enophthalmos), the pupil reacting normally to light and accommodation.

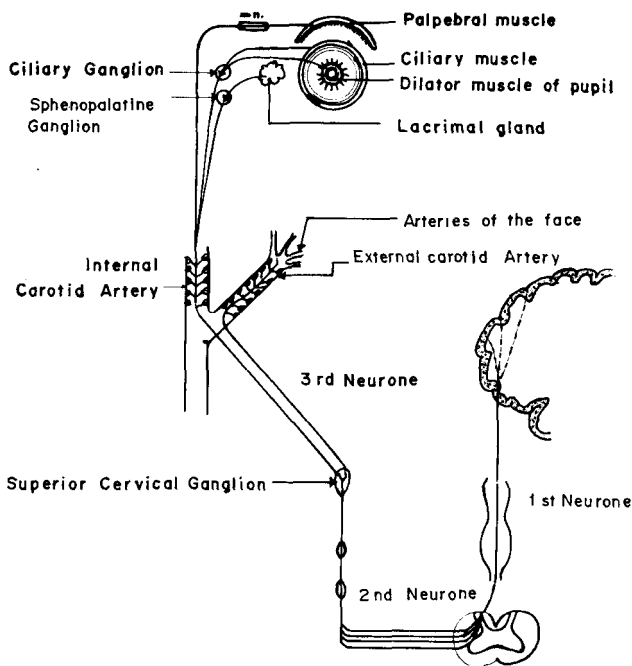


Fig. 2.6 : Sympathetic pathway

Horner's syndrome can result from lesions of the brainstem such as multiple sclerosis, or pontine glioma. Lesions of the cervical spinal cord such as syringomyelia and cranio-vertebral anomalies are other causes of Horner's syndrome. Carcinoma of the apex of the lung Pancoast's tumour, and cervical rib cause Horner's syndrome by involvement of the cervical sympathetic pathways. Neoplastic infiltration, and tuberculous lymph adenitis sometimes lead to Horner's syndrome. It is possible to differentiate lesions of the first and second order neurons from the third order neurones by the cocaine and hydroxyamphetamine test. When a drop of cocaine is put in either eye, the pupil will dilate poorly to cocaine no matter what the level of the lesion. The cocaine test confirms or excludes the presence of a Horner's syndrome. The hydroxyamphetamine test involves the instillation of 1% hydroxyamphetamine. Damage to first or second order neurons results in pupillary dilatation. When third order neurones are affected no dilatation occurs. Pupillary dilation depends upon the ability of the third order neurones to produce, transport, and store norepinephrine. When hydroxyamphetamine is instilled, the pupils dilate because norepinephrine is released. When third order neurones are involved, norepinephrine production and

storage is affected. Hence the pupil does not dilate on instillation of hydroxyamphetamine. The cocaine-hydroxyamphetamine test therefore helps to localise the lesion to the first, second, or third order neurones of the sympathetic pathway.

Nystagmus Nystagmus is described at this time because it is generally noted whilst examining eye movements, though the lesions that cause nystagmus are usually located in the vestibular pathways. Nystagmus is a disturbance of ocular posture which is characterised by oscillatory movements of the eyes. Nystagmus may be horizontal, vertical or rotary. There is a tendency for the laterally deviated eye to drift slowly to the midline, with a fast flicker of movement to the original position. The nystagmus is designated after the direction of the fast component, for instance if the fast component is to the right the nystagmus is called a right nystagmus. However, it is the slow drift of the eyes which is secondary to the pathologic lesion, the fast component is the corrective movement undertaken to regain the original position.

Nystagmus is divided into three degrees. First degree or minor nystagmus is present only when the eyes are deviated to the extremes of lateral gaze. Second degree nystagmus is apparent with minimal deviation of the eyes and third degree nystagmus is seen when the eyes are in the resting position. 'Pendular' nystagmus is seen in ocular lesions with diminished visual acuity such as optic atrophy, albinism, congenital cataracts, and total colour blindness. Congenital nystagmus is also pendular in type and is present from birth. It may be familial, the movements being gross, and associated with head nodding. This type of nystagmus should be distinguished from all other types of pendular nystagmus.

Nystagmus may occur due to lesions anywhere along the vestibular pathways. With chronic lesions of the cochlea such as Meniere's disease, and in tumours of the vestibular nerve such as acoustic neuroma, no nystagmus may be seen if central compensation has occurred. Nystagmus is marked in acute lesions such as labyrinthitis or neuronitis, till central compensatory mechanisms have been established. As brainstem lesions affect these compensatory mechanisms, nystagmus is always seen with lesions of the brainstem. The nystagmus may be horizontal, vertical, or rotary. Vertical nystagmus usually indicates an intrinsic lesion of the brainstem, such as multiple sclerosis, a glioma, or an infarct. The left vestibular pathways move the eyes to the right. Hence the eyes will tend

to drift towards the left with a lesion on this side. A corrective fast movement occurs to the right, therefore, left vestibular lesions produce right nystagmus.

Cerebellar lesions result in nystagmus towards the diseased side, which is due to involvement of the vestibular connections of the cerebellum. Some cases of cerebellar nystagmus require closing the eyes, or examining the patient in a dark room, before the nystagmus is observed. A new technique that records nystagmus is known as electronystagmography.

Nystagmus occurs with barbiturate or phenytoin intake, and in Wernicke's encephalopathy. Downbeat nystagmus occurs with lesions of the cervico-medullary junction such as the Chiari malformation.

In internuclear ophthalmoplegia, the lesion is located in the medial longitudinal fasciculus. There is failure of adduction on lateral gaze and nystagmus may be seen in the abducting eye. Internuclear ophthalmoplegia is seen in intrinsic lesions of the brainstem and is commonly seen in multiple sclerosis.

Trigeminal nerve (Fifth cranial nerve) It is a mixed nerve, conveying sensation to the face and providing the motor supply to the muscles of mastication. Sensory fibres from the face terminate in the Gasserian ganglion. Second order neurone fibres relay to the brainstem, from where they take diverse courses. Fibres carrying tactile, and discriminatory tactile sensations from the face, course cephalad from the primary sensory nucleus in the pons to the midbrain. Fibres carrying thermal and pain sensation from the descending tract of the trigeminal nerve and end in the elongated nucleus which lies by the side of the tract. The tract and nucleus descend to the level of the first cervical segment of the spinal cord. The fibres are arranged so that fibres carrying sensation from the upper face lie in the lowest part of the tract, and those from the lower face lie in the uppermost portion of the tract. The mesencephalic nucleus of the fifth nerve receives proprioceptive impulses from the muscles of mastication.

The motor fibres originate from the nucleus which lies medial to the large sensory nucleus in the pons. The nerve emerges from the pons as two closely approximated roots, a comparatively small motor root and a larger sensory root. The nerve pierces the dura at the apex of the

petrous bone to form the Gasserian ganglion. The trigeminal nerve has three divisions, the ophthalmic, maxillary, and mandibular. The ophthalmic division supplies the conjunctiva, the lacrimal gland, the skin of the nose, scalp, forehead, and the upper eyelid. The maxillary division supplies the skin of the lower eyelid, most of the upper lip, the cheek, temporal area, and the nasal mucosa, pharynx, soft palate and tonsils. The mandibular division supplies the lower part of the face, the lower lip, tongue, the lower teeth, and the salivary glands. The motor root travels with the mandibular division to supply the masseter, temporalis and pterygoid muscles.

Facial sensation is checked with a wisp of cotton first. The corneal reflex is checked by twisting the cotton wisp to a point and touching the lower and outer portion of the cornea. The patient is asked to look upwards and inwards. If the corneal reflex is present both eyes will blink simultaneously. The afferent arc of the corneal reflex is via the ophthalmic division of the fifth, and the efferent arc is via the seventh nerve, which supplies the orbicularis oculi. Absence of the corneal reflex on the side may occur secondary to a lesion of the fifth or seventh nerve. If the fifth nerve is involved, there will be no response from *either* lid when the abnormal side is stimulated, and a normal response from *both* lids when the normal side is stimulated. If, on the other hand, the seventh nerve is involved, there is no response on the side of the facial paralysis, irrespective of the side being stimulated.

The pterygoid muscles open the jaw and simultaneously pull it forwards. Weakness of the pterygoids results in deviation of the jaw to the weaker side on opening the mouth. This jaw deviation is best observed by keeping the relative position of the central incisor teeth as a guide, or by keeping one finger in the midline under the chin and asking the patient to open his mouth slowly. Remember the jaw deviates to the *side* of the lesion. Function of the masseter and temporalis muscles is tested by asking the patient to bite hard and appreciating the contraction of the muscles on palpation.

Trigeminal neuralgia This extremely painful condition affects one or all three divisions of the trigeminal nerve. The second and third divisions are the most commonly affected. Women are affected twice as commonly as men. The disease is seen in our country after the age of forty. There is no known cause. Pain occurs in paroxysms lasting one to two minutes. The attacks are described as agonising, the pain being

sharp and lancinating. The patient is free of pain between attacks. Since facial movement may trigger the attacks, the patient appears very apprehensive and is afraid to move his facial muscles. Attacks may be precipitated by speaking, eating or even by a blast of air on the face. There are often "trigger" points on the face which, if touched, may set off an attack. The disease generally has a relapsing and remitting course. Each relapse lasts for a longer time, the periods of remission getting shorter. In 'Trigeminal Neuralgia' no sensory or motor deficit can be demonstrated on examination of the areas supplied by the trigeminal nerve. The corneal reflex is normal. If a deficit is present, a tumour or vascular malformation should be suspected and appropriate investigations undertaken. Trigeminal neuralgia is also known as "tic douloureux" because muscle twitches accompany pain during attack.

Carbamazepine (Mazetol) is very effective in controlling the pain. 100 to 200 mgms given as a single dose affords relief for a few hours, the same dose being repeated according to the patient's requirements. Diphenylhydantoin in a dose that varies between 100 and 300 mgms may be added to the regimen. If the pain proves intractable to medical therapy, alcohol injection of the Gasserian ganglion or surgical sectioning of the nerve root should be considered. Heat coagulation of the root by using radio frequency has been tried successfully and may become the treatment of choice.

Facial Nerve (Seventh Cranial Nerve) The facial nerve is a motor nerve that together with its sensory-parasympathetic component, the nervus-intermedius emerges from the brainstem in the region of the cerebello-pontine angle. The motor root arises from its nucleus situated in the rostro-lateral part of the pons. The nerve, in its intrapontine course, winds around the nucleus of the abducens nerve and emerges from the ventral surface of the pons with the nervus intermedius. It lies in close proximity to the eighth nerve as it enters the internal auditory meatus. Within the auditory canal the facial nerve and the nervus intermedius continue in their canal, the facial canal or the fallopian canal, giving off the branch to the stapedius muscle. The chorda tympani, a branch of the facial nerve is given off just before the facial nerve leaves its canal. The chorda tympani now travels with the lingual nerve. The facial nerve emerges through the stylomastoid foramen, travels through the parotid gland and divides into a number of branches that supply all the muscles of the face, the frontalis, and the platysma. The secreto-motor fibres innervate the lachrymal and salivary glands.

The chorda tympani carries the taste fibres from the anterior two-thirds of the tongue.

Facial paralysis results in a widening of the palpebral fissure on the affected side, the naso-labial fold is obliterated and there is drooping of the angle of the mouth. However, a very useful sign, detectable even with minimal involvement of the facial nerve, is an absence of blinking on the side of the lesion. As the facial nerve controls facial musculature, testing its function involves the testing of facial movement. The patient is asked to frown and wrinkle the forehead, to test the function of the frontalis muscle. Next, she is asked to close her eyes tightly, burrowing her eyelashes deeply. In a case of facial paralysis, the eye may not close at all, or, if the weakness is minimal, the eyelashes may not be burrowed in deeply. Normally, eye closure, results in the eyeballs rolling upwards, and slightly outwards. In facial palsy, the eyeball is *seen* to roll up as the eye cannot be closed due to weakness of the orbicularis oculi. This phenomenon is known as "Bell's phenomenon".

The patient is then asked to show the teeth, and say 'eeeeee'. There is deviation of the mouth to the *normal* side, with obliteration of the nasolabial fold on the paralysed side.

As mentioned earlier the sensation of taste is carried by the chorda tympani. The taste fibres end in the tractus solitarius in the pons. Taste sensation from the posterior one-third of the tongue is carried by the glossopharyngeal nerve to the tractus solitarius. From here, relay fibres cross the midline and proceed rostrally with the posterior longitudinal bundle as the gustatory fillet. They end in the hypothalamus. The sensation of taste is tested by first asking the patient to protrude the tongue. Wipe the tongue dry, and put a drop of a sweet or salty substance on the tongue. The patient then is asked to put the tongue back in the mouth and not to swallow till he or she has recognised the taste.

In some instances of facial weakness, there may be a weakness of limb musculature on the same or the opposite side depending on the level of the lesion. These instances will be discussed more fully in the section on the motor system.

The table below differentiates between infranuclear and supranuclear lesions of the facial nerve. It also lists the deficits that result from lesions of the facial nerve at various points along its course.

TABLE 2.9

Infranuclear :

a) At stylomastoid foramen and distal to it.	Paralysis of facial musculature only
b) In the facial canal before branching of chorda tympani	Paralysis of facial musculature and loss of taste anterior two-thirds of tongue.
c) In the facial canal before branching of the nerve to stapedius.	Paralysis of the facial musculature, loss of taste sensation as above and hyperacusis.
d) In the posterior fossa.	Eighth nerve or other lower cranial nerves simultaneously involved or bilateral facial paralysis as in Guillain-Barre' syndrome.

Nuclear

Sixth nerve simultaneously involved and other long tract involvement.

The facial nerve is commonly affected in tumours of the cerebello-pontine angle such as acoustic neuroma, basal meningitis, sarcoidosis, otitis, parotitis, the Guillain-Barre syndrome, trauma, and leukemia.

The commonest cause of facial palsy is "Bell's palsy" a lesion with no known cause, but where a viral aetiology is nevertheless suspected. It can occur at any age, though it is uncommon in children, and its incidence increases upto the age of forty. There is no predilection for either sex. The onset is usually acute, total paralysis developing in a day or two. Many patients complain of pain behind the ear, for a few days before and after the onset of paralysis. Taste fibres are involved in a small number of patients, whilst many complain of excessive tearing. Hyperacusis occurs when the nerve to the stapedius is also affected. The normal function of the stapedius is to reduce the amplitude of vibratory excursions of the stapes in the oval window. Hyperacusis is an increased acuity of hearing and undue sensitivity to low tones. Unilateral facial weakness is partial in 25% and total in 75% of the patients. 80% of the patients recover rapidly and fully. Steroids given early in the illness may hasten the recovery. 60 - 80 mgms of prednisolone is given for a week and then tapered rapidly. Physiotherapy helps the patients to deal with the disability but does not speed the recovery. In cases with incomplete or partial recovery, electromyography helps in determining

prognosis. The following guidelines are useful to assess prognosis : younger patients with partial paralysis, preservation of taste, good submandibular flow, and normal conduction on electromyography, will recover fully.

The acoustic nerve (Eight cranial nerve) It consists of two separate divisions carrying the sensation of hearing and balance. The vestibular division arises from the large bipolar cells of the vestibular or Scarpa's ganglion, situated at the distal end of the auditory canal. It travels in the canal with the cochlear division, enters the brainstem behind the facial nerve and ends in the four vestibular nuclei. Fibres from these nuclei establish connections with the nuclei of the third, fourth, and sixth cranial nerves, the cerebellum, the reticular substance, and centre in the temporal lobes.

The cochlear division of the eighth nerve originates in the bipolar cells of the spiral ganglion in the bony canal of the cochlea, and is associated with the hair cells of the auditory receptor organ. The nerve accompanies the vestibular division and enters the brainstem close to the inferior cerebellar peduncle, lying lateral and inferior to the vestibular branch. On entering the brainstem the nerve divides into two branches and ends in two cochlear nuclei. Second order fibers from these nuclei ascend to the auditory centres in the temporal lobes.

The function of the cochlear division is tested clinically by checking the patient's ability to hear normally as well as whispered speech in each ear. The Rinne's and Weber's tests help determine whether defective hearing is due to a disease of the inner ear, or of the cochlear nerve. A normal person has better conduction of sound through air than through bone. In Rinne's test, the external auditory canal is blocked with a finger, and then a vibrating tuning fork which has a frequency of 512 is placed over the mastoid. The patient is asked to indicate when he stops hearing the sound. The tuning fork is now removed from the mastoid and held in front of the ear. A normal person will still perceive the sound while a person with middle ear disease will not. This is due to bone conduction of sound being better than air conduction in middle ear disease. Also the vibrating tuning fork is heard better in the affected ear, when the fork is placed in the middle of the forehead. The Weber's test is said to lateralise to the affected ear. Hearing can be further checked by the use of sophisticated tests such as audiometry and loudness recruitment. The acoustic reflex and electrocochleography are

other tests that are used in the investigation of deafness.

Dysfunction of the vestibular division gives rise to vertigo. Although not a part of routine neurological examination, tests of vestibular function involve alteration of the flow of current in the endolymph within the semicircular canals. *The Caloric tests* Each ear is tested separately. The patient lies comfortably with the head flexed by 30 degrees so that the horizontal canal lies in the vertical plane. Each ear is irrigated with either 250 ml of cold water at 33 degree celsius or with hot water at 44 degree celsius for 40 seconds. The patient is asked to look straight ahead whilst the ear is being irrigated. The patient has a subjective feeling of vertigo, and the eyes demonstrate nystagmus to the opposite side if cold water is used and to the same side if hot water is used. Nystagmus normally lasts for about two minutes. Electronystagmography is a test which records nystagmus and may help in differentiating peripheral from central nystagmus.

In Barany's rotation test nystagmus is produced by rapidly rotating a person who is seated in a special chair. Ten revolutions are completed in 20 seconds and the chair is brought to an abrupt standstill. Nystagmus occurs in the direction opposite to the direction of the rotation. If there is labyrinthine dysfunction in that direction, nystagmus is markedly reduced.

The causes of vertigo have already been detailed in chapter one. The causes of deafness are varied and include *inherited* causes such as Refsum's disease or hereditary sensory neuropathy. *Haemolytic disease of newborn* and *anoxia* at birth affect the auditory pathway and the cochlear nuclei. *Head injury* commonly produces vertigo, and rarely deafness. *Meningitis* particularly tuberculous meningitis, and meningococcal meningitis in children often lead to deafness. *Viral infections* can cause either deafness or vertigo. *Drugs* that damage the eighth nerve include quinine, the salicylates, streptomycin, kanamycin, ethacrynic acid and frusemide. Tobacco and alcohol have been reported as causing eighth nerve damage. *Hyperthyroidism* and *diabetes* are common metabolic causes whilst *vascular* lesions of the brainstem, especially a lesion in the anterior inferior cerebellar artery leads to unilateral loss of hearing. *Tumours of the cerebello-pontine angles* result in gradual hearing loss.

Meniere's disease It is characterised by paroxysmal attacks of severe vertigo, and is thought to be due to vasospasm and pathologic

dilatation of the endolymphatic system in the cochlea. It is commoner in males than in females, and is often associated with migraine. It may present at any age, the incidence progressively increasing after the age of thirty. The onset of vertigo is sudden and severe, the attack being accompanied by a sensation of fullness in the ear, and systemic complaints like nausea, vomiting, sweating and pallor. Tinnitus and deafness occur, nystagmus is seen, and the attack lasts for a few minutes to a few hours. Recurrent attacks are infrequent in the initial course of the disease, and may occur every few weeks or even after many months. As the frequency of attacks increases, there is increasing deafness, with vertigo ceasing at the onset of total deafness. Early in the disease, the patient is unable to tolerate high pitched sounds, which accounts for the "recruitment phenomenon". An individual attack is treated with sedatives and triflupromazine hydrochloride, Phenobarbital and vasodilators like nicotinic acid may help in reducing the frequency of the attacks. In severe cases, cervical sympthectomy or surgical destruction of the labyrinth may be necessary.

Acoustic neuroma The tumour arises in the vestibular division of the eighth nerve and grows very slowly, Bilateral acoustic neuromas are almost always seen in patients with neurofibromatosis. Tinnitus is usually the first symptom, followed by a gradual diminution of hearing. Facial palsy occurs very late in the course of the disease, and is rare in spite of the close proximity of the facial nerve to the eighth nerve. The tumour compresses the fifth nerve leading to loss of the corneal reflex on the affected side. Numbness over the face may be present, though total loss of sensation is rare. There is generally no pain in the trigeminal distribution. Further tumour growth results in pressure on the brainstem. Ataxia, spasticity, hyperreflexia, and an extensor plantar response may be present at this stage. By the time papilloedema has developed, the tumour is fairly large in size. Plain x-rays of the skull reveal widening of the internal auditory meatus. Other investigative procedures include audiometry, electronystagmography, a study of brainstem auditory evoked potentials, and metrizamide computerised tomography. Treatment consists in surgical removal.

Glossopharyngeal nerve (Ninth Cranial Nerve) Though a mixed nerve, its motor supply is only to the stylopharyngeus muscle. It also carries taste fibres for the posterior one-third of the tongue. The nerve enters the skull through the jugular foramen, and enters the brainstem, lateral to the medullary olives. The fibres end in a nucleus, in the

medulla, in the floor of the fourth ventricle. The afferent arc of the gag reflex is carried via the ninth nerve, the efferent via the vagus (tenth) nerve. When the gag reflex is impaired sensation over the palate and posterior pharyngeal wall has to be tested with a long hat pin. Glossopharyngeal neuralgia is a rare disease that occurs after the age of thirty. The onset is sudden, the attacks characterised by shooting, agonising pain on one side of the palate and pharynx. The attacks are readily provoked by swallowing, and sometimes may only be controlled by sectioning of the nerve.

Vagus nerve (Tenth Cranial Nerve) It is a mixed nerve, arising in the medulla from the nucleus ambiguus and tractus solitarius. It leaves the skull through the jugular foramen with the ninth and eleventh nerves. The auricular branch supplies the concha of the external ear, the pharyngeal branch supplies the levator of the palate, the constrictors of the pharynx, the cricothyroid muscle, and sensory fibres to the larynx. On the right, the recurrent laryngeal nerve arises at the root of the neck and on the left, it arises in the upper thorax. The recurrent laryngeal nerve supplies all the muscles of the larynx except for the cricothyroid. The vagus nerve carries visceral afferent and efferent fibres for all the thoracic and abdominal organs.

Examination of the tenth nerve includes observing palatal movements, the intonation of speech, and the ability to cough. Bilateral palatal weakness results in a low lying palate, a nasal twang, and nasal regurgitation. Unilateral weakness on the other hand, results in the palate being pulled to the normal side when elevated. It is best observed when the median raphe of the palate is seen to move over to one side instead of going up in midline. Unilateral vocal cord paralysis leads to hoarseness of the voice, the cord lies immobile in the midline, and the patient is unable to cough normally. A hacking or bovine cough is produced instead.

Lesions that produce ninth and tenth nerve paralysis can be divided into two groups: intramedullary lesions that cause nuclear and supra-nuclear palsies, and extramedullary lesions that can occur either intra- or extra-cranially. The (Table 2.10) lists the causes of ninth and tenth nerve lesions.

TABLE 2.10

LESIONS CAUSING NINTH, TENTH AND ELEVENTH CRANIAL NERVE PALSIES.

Intramedullary :

Lesions	Unilateral or bilateral involvement	Effect
Pontine tumours	Bilateral	Palatal, pharyngeal Laryngeal palsy.
Syringobulbia	Bilateral	As above
Motor Neurone disease	Bilateral	As above, sensations preserved.
Poliomyelitis	Unilateral or bilateral	Can affect any one or all three, sensations preserved.

Extramedullary, Intracranial

Cerebello-Pontine angle Tumours.	Unilateral	Gradually progressive palatal, pharyngeal and Laryngeal paralysis.
Meningitis	Unilateral or Bilateral	As above.
Guillain-Barre syndrome (Polyneuritis cranialis)	Bilateral	Palatal, pharyngeal and/or laryngeal.
Diphtheretic neuritis	Unilateral or bilateral	Mainly palate.

Extracranial

Jugular foramen syndromes	Unilateral	IX and XI, or X and XI or X, XI and XII
Neurofibroma, Meningioma, Glomus tumours.		with Horner's syndrome
Recurrent laryngeal palsy intrathoracic tumours	Unilateral	
Aneurysm of aorta.		
Left atrial enlargement		
Thyroid tumours		
Neoplastic and Kock's lesions of cervical and mediastinal glands	Unilateral	Laryngeal palsy
Operative lesions.		Hoarse voice

Accessory Nerve (Eleventh Cranial Nerve) It is a pure motor nerve consisting of two parts, a medullary and a spinal part. The medullary portion arises from the nucleus ambiguus and the spinal portion from the upper three cervical segments. The spinal portion emerges between the ventral and dorsal roots of the cord, and forms a trunk that ascends through the foramen magnum to join the medullary portion. This common trunk then leaves the skull through the jugular foramen with the ninth and tenth nerves. It supplies the sternomastoid and trapezius muscles. Lesions of the accessory nerve result in wasting and weakness of the sternomastoid and the upper part of the trapezius. The shoulder droops on the side of the paralysed trapezius, the scapula falling away laterally from the thoracic cage, the superior angle being displaced to a greater degree than the inferior angle. Power in the trapezius is tested by asking the patient to shrug his shoulders upwards against resistance. The sternomastoid is tested by asking the patient to turn the head to one side, while the examiner applies resistance at the chin. In cases of bilateral weakness of the sternomastoid, the patient is asked to lift the head off the bed against gravity.

Hypoglossal nerve (Twelfth Cranial nerve) It is also a pure motor nerve, which supplies the muscles of the tongue—the hyoglossus, styloglossus, genioglossus, and geniohyoid muscles. The nerve originates in the hypoglossal nucleus which lies in the posteromedial part of the medulla, beneath the floor of the fourth ventricle. It pierces the dura, enters the hypoglossal canal, and on leaving the skull passes vertically downward to the level of the angle of the mandible and reaches the tongue.

Lesions of the hypoglossal nerve lead to atrophy and paralysis of the tongue on the side of the lesion. Fasciculations may be seen when the lesion affects the lower motor neurone. The tongue seems to be full of contracting muscle fibres as it lies in the mouth. On protruding the tongue deviates to the paralysed side. Due to the close proximity of the last four cranial nerves, lesions (see table II, 10) tend to affect them in varying combinations. The presenting symptoms remain the same irrespective of the cause. These symptoms include hoarseness of the voice, dysphagia and nasal speech. Bulbar palsy means paralysis of the medullary region, and is seen in poliomyelitis.

CHAPTER - III

MOTOR SYSTEM

The motor system consists of the motor neurone and their axons together with the effector organs, the muscles. Those neurones located within the cerebrum are grouped under upper motor neurone while those in the spinal cord are called lower motor neurone. Their activity is further modulated and integrated by extrapyramidal, cerebellar and vestibular influences. The origin and course of the pyramidal tract is shown in the figure 3.1.

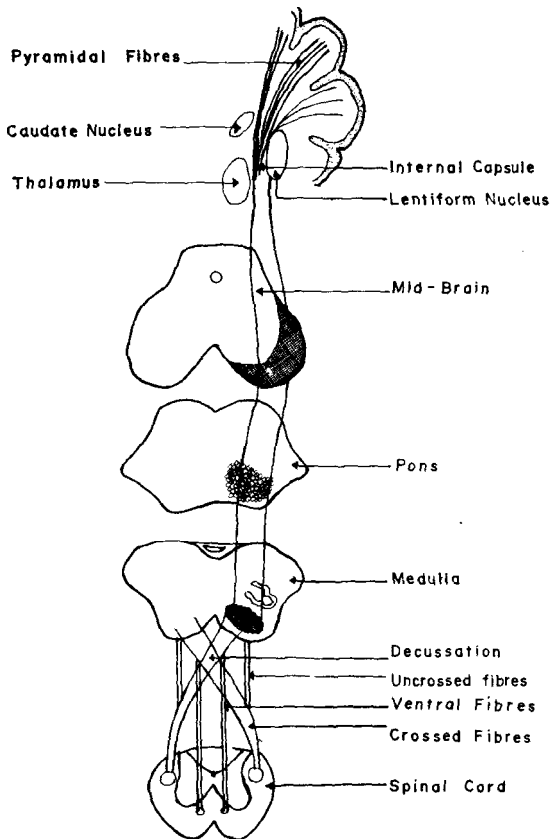


Fig. 3.1 : Schematic diagram showing corticospinal pathways

MUSCLE TONE The resistance felt on passive movement of a joint is known as 'tone'. Current theories explaining this phenomenon emphasise the role of mechanical properties of the muscles, joints and ligaments. Earlier concepts which emphasised continuous contraction of muscles are no longer accepted. The anatomical basis of tone is the stretch reflex arc which receives afferent impulses from the muscle spindles and Golgi tendon organs. Efferent impulses from the alpha motor neurone govern the activity of extrafusal fibres. The stretch reflex is facilitated by the vestibular and cerebellar systems, while the reticulospinal, **extrapyramidal**, and pyramidal systems exert an excitatory influence on the arc.

Tone is tested by passively moving different joints with both peripheral and axial joints being tested. The resistance offered during these movements is assessed as normal, increased or decreased in tone. We have already discussed disturbances of tone in upper and lower motor neurone lesions in the previous chapter, we will only remind the reader that spasticity occurs in chronic upper motor neurone lesions, and flaccidity occurs in both 'neural' and 'spinal' shock.

Spasticity On passive flexion or extension of a limb increased resistance is felt at the beginning, with a 'give' as the movement continues. It can be compared to the opening of the penknife hence the name 'clasp knife' rigidity.

Rigidity Tone is increased in both the agonists and antagonists so that passive movement of a joint gives the impression of a lead pipe is being bent, hence the name 'lead pipe' rigidity for this disturbance of tone. Lead pipe rigidity is characteristic of disease of the basal ganglia.

Cog-wheel rigidity A patient with Parkinson's disease has an increase in the tone that is described as 'cog-wheel' rigidity. On moving the wrist through full range of extension, the flexors alternately offer resistance and give, akin to the movement of a cog-wheel. Cog-wheel rigidity is best demonstrated by steady slow movements of flexion and extension at the wrist, and pronation and supination at the elbow.

Myotonia It is demonstrated by asking the patient to grip the examiner's hand forcefully, and then to let go of the hand suddenly. A patient with myotonia is unable to release the grip suddenly due to continued after contraction of the long flexors of the fingers. Cases with myotonia are rare in India. Myotonia can result from disease of muscle

or of the peripheral nerve. Percussion myotonia is demonstrated by tapping the thenar eminence with a reflex hammer and observing whether a 'dimple' appears. The 'dimple' is due to the sustained contraction of a few muscle fibres underneath the site of the tapping. Myotonia is seen in myotonia congenita, and myotonic dystrophy.

Hypotonia Hypotonia is seen in peripheral nerve and cerebellar lesions. In extreme hypotonic state the muscles feel flabby and the limb is flail. The joints can be moved to wider range of movements, like that of a 'Rag doll'. Assessment of hypotonia is subjective and minimal diminution of tone is difficult to assess. The child or infant with hypotonia lies with lower limbs kept like those of a frog (Frog-leg position).

Motor power is assessed by asking the patient to contract his muscles against resistance offered by the examiner, or by asking the patient to hold a joint in a particular position, and the examiner then attempts to move the joint in the opposite direction. If there is considerable weakness, the patient may be unable to lift a limb against gravity. Motor power is then tested by eliminating gravity, and testing muscle strength. Power should be graded according to the table below.

TABLE 3.1

Scale used to grade motor strength

Full power	5
Minimal weakness	4
Movement against gravity but not against resistance	3
Movement with gravity eliminated	2
Flicker of movement	1
No movement	0

The most striking degenerative diseases involving the motor system are amyotrophic lateral sclerosis, the progressive bulbar palsy and the motor neurone disease. In these three distinct clinical syndromes, there is varying degree of degenerative process involving both the upper and lower motor neurones. These three syndromes can be grouped under a single encompassing term 'Motor neurone disease'.

MOTOR NEURONE DISEASE

The disease occurs sporadically in India. However, familial cases are known to occur in certain parts of the world, specially in the islands of Guam in the Pacific, affecting the local Chamorros population. The disease usually is of adult life beginning after the age of forty, a milder form occasionally occurring in younger individuals. There is a male predominance except in progressive bulbar palsy where both sexes are equally represented.

The pathogenesis of the disease still remains obscure. Heavy metal toxicity, virus infection and even trauma have been blamed at one time or the other. An occasional case of the disease is known to occur following the attack of poliomyelitis or encephalitis. The lateral corticospinal tracts undergo degeneration starting from the centre, and chromatolysis of cells occur in the nuclei of the lower cranial nerves and the anterior horn cells of the spinal cord. The 3rd, 4th and 6th cranial nerve nuclei are spared.

Progressive bulbar palsy The initial symptom may either be dysphagia or dysarthria. The dysarthria is due to involvement of the tongue, perioral muscles and the soft palate. The voice starts to acquire a nasal quality with poor articulation of the linguals and gutturals. The food sticks in the throat, at times causing choking and the liquids regurgitate through the nose. This involvement of the labio-glosso-pharyngeal muscles is progressive.

On examination facial weakness is demonstrated, the face is drooping, the mouth remaining open with saliva dripping out. The tongue is atrophic, small and wrinkled and shows profuse fibrillations. The palate hangs low in the pharynx and is immobile, however, the gag reflex may be preserved. Fasciculations may be seen in the muscles of the shoulder. Signs of early pyramidal involvement may be present and the jaw jerk may be brisk.

The disease progresses relentlessly necessitating tube feeding and tracheostomy in the later stages. Patients usually die due to respiratory infection, within 12 to 18 months after the onset of the disease.

Amyotrophic lateral sclerosis In this form of the disease, the initial symptoms is difficulty in walking due to spasticity of lower limbs. The patient may complain of heaviness or dragging of one or both feet. The

examination at this stage will reveal mild spasticity with brisk reflexes in all four limbs and an extensor plantar response. The abdominal and the cremasteric reflexes may be preserved even exaggerated (dancing umbilicus). Sparse fasciculations can be observed or induced by tapping in the muscles of the shoulder, quadriceps and calves. There is no sensory involvement and the sphincters remain intact till last. Pseudobulbar and bulbar palsy occurs within two or three years of the onset of the disease bringing an end to patient's life.

Progressive muscular atrophy The disease usually commences by affecting the small muscles of the hands. It begins in one hand, ascending upwards to involve the forearm, arm and shoulder muscles. It rapidly involves the other hand. Fine movements of the fingers specially the thumb are initially affected. It is difficult to oppose the thumb producing difficulty in holding a pen or small objects. As the atrophy progresses, all the small muscles of hand are involved, the grip becomes weak and claw shaped deformity of the hand is noted. Very rarely the disease may begin in the lower limbs with patients having difficulty to wear and maintain chappals, in the foot affected. The diagnosis may be difficult at this stage and plexitis or spondylosis may be suspected. Electromyographic examination at this stage even may be helpful. The clinical examination may reveal sparse fasciculations in the shoulder and lower limb muscles. The tendon reflexes in the atrophic limb are lost with normal or brisk reflexes in the lower limbs. Plantar reflex becomes extensor in the course of the disease. Sensations and sphincters remain intact. The progress and the course of the disease is variable. Eventually the bulbar muscles are involved and the patient survives for 5 to 15 years after the onset of the disease. Electromyographic examination is the most useful investigation in establishment of the diagnosis. The treatment is symptomatic.

Many Greek and Latin terms are used in Neurology to describe motor deficits. We have already defined the terms hemiplegia, paraplegia, quadriplegia, and diplegia in chapter 1. We shall now discuss the effects of these deficits and the lesions responsible for causing them in greater detail.

HEMIPLEGIA The commonest cause for a hemiplegia is a vascular lesion involving the corticospinal tract from its origin in the cerebral cortex, to its decussation in the medullary pyramids. *Apoplexy*, a term known to physicians through the ages, is used specially for intracerebral haemorrhage.

An acute lesion of the cortico-spinal tract results in a state of flaccid paralysis due to "neural shock". Flaccid, means total loss of tone, and the upper and lower limbs are markedly hypotonic or even atonic. The "shock" clears in two to three weeks and spasticity starts to develop. The signs that develop are due to two main factors. These were described by Hughlings Jackson as 'negative' elements which are due to loss of function of the pyramidal tracts, (paralysis), and 'positive' elements (spasticity, exaggerated reflexes), that result from the loss of the inhibitory influence exerted by the pyramidal tract. Systems that are intact are 'released' and now exert their influence.

Negative signs An acute lesion of the right hemisphere that affects the pyramidal tract results in a weakness of conjugate deviation of the eyes to the left side. The eyes 'look' at the lesion, due to the unopposed action of the intact left hemisphere. This ocular deviation is not seen after a few days of the insult, and is therefore not observed in hemiplegia of long standing. Since head movement is closely related to eye movement, the head is also deviated to the side of the lesion at the onset of the hemiplegia.

Movements of the upper part of the face are bilaterally synchronous and are controlled by both hemispheres. Hence the upper face is spared in a unilateral lesion involving the hemispheres. The lower face is however, paralysed and there is flattening of the nasolabial fold. The patient is unable to purse his lips. Facial movements that occur because of emotional changes, such as laughing or crying, remain intact because fibres subserving these movements do not travel with the pyramidal tracts. Sometimes jaw and tongue deviation is seen transiently.

As the effects of the 'neural shock' subside, motor tone increases, gross limb movements may recover, but fine skilled movements such as those involving the fingers suffer the most and never recover fully. Power first returns to the flexors of the upper limb and extensors of the lower limbs. The typical hemiplegic posture results from a dominant flexor tone in the upper limbs and extensor tone in the lower limbs. The posture, when fully developed shows the arm adducted at the shoulder, flexed at the elbow, wrist, and fingers, with the thumb opposed to the palm. The leg is externally rotated, extended at the knee, and the foot plantar flexed. Minimal pyramidal involvement becomes evident due to difficulty with finger movements as in buttoning, and writing. Hemiplegic gait involves circumduction of the foot, to avoid scraping the floor because of the extended knee and the footdrop.

Positive symptoms As the effects of the 'neural shock' wear off spasticity develops. Tone returns first to the flexors of the upper limb and extensors of the lower limb. The deep tendon reflexes which were absent during the early period, become exaggerated, with clonus at the wrist and ankle. In cases with severe hypertonia, patellar and finger clonus may be demonstrated. The abdominal and cremasteric reflexes are absent, and the plantar response is extensor. Hoffman's reflex may be elicited; it consists of flexion of the fingers, on sudden flexion by flicking of the middle finger at the distal interphalangeal joint.

LOCALISATION OF THE LESION

As mentioned earlier, hemiplegia can result from a lesion of the pyramidal tract anywhere from its origin to its decussation. Associated symptoms and signs help to localise the lesion at various sites in the cortex and brainstem.

In a cortical lesion, the paralysis always remains flaccid. Focal convulsions may herald the onset, and there is generally paralysis in only one limb. This is due to the extensive origin of the pyramidal tract from the cortex. A lesion in the internal capsule on the other hand, leaves a profound hemiplegia with a sensory deficit, and hemianopia, because these pathways are crowded together in this region.

LESIONS INVOLVING THE TERRITORY OF THE MIDDLE CEREBRAL ARTERY

The middle cerebral artery arises at the bifurcation of the internal carotid artery above the cavernous sinus. The artery then enters the Sylvian fissure where it divides into three branches, the precentral, posterior parietal, and posterior temporal branches. Also given off in the sylvian fissure are six to ten thin penetrating vessels known as lenticulo-striate branches that supply the basal ganglia, internal capsule and the optic radiation.

Occlusion of the main trunk It causes a large area of infarction, and massive oedema of the affected hemisphere. The oedema may be so severe with pressure on the brainstem causing coma. Occlusion of the middle cerebral artery on the dominant side produces global aphasia, and apraxia whilst non-dominant lesions produce loss of body image on the contralateral side, and denial of the existence of the left side of the body. There is a profound flaccid hemiplegia, the arm being more affected than the leg. Hemianaesthesia and a complete hemianopia also

occur in this most devastating stroke. Death occurs if a pressure cone forms and gives rise to severe brainstem compression.

Occlusion of the lenticulo-striate branches It usually leads to a pure motor hemiplegia, with flaccidity occurring only fleetingly in the early stage of the disease. Tone begins to recover within a few hours, and exaggerated reflexes and an extensor plantar response are also observed. Speech may be slurred due to facial weakness, but there is no aphasia. Power recovers gradually, and the patient is left with the typical hemiplegic posture and gait. This is the commonest kind of stroke and has a fairly good prognosis for recovery.

Occlusion of the terminal branches Dominant hemisphere lesion :
Occlusion of the precentral branch : This vessel supplies Broca's area and the motor area for the face and arm. There is paralysis of the face and arm and a total inability to speak, though the ability to comprehend spoken speech is retained. The patient is unable to write, and appears markedly frustrated by the speech deficit. The chances of recovering speech are poor, though speech therapy can help to some extent. Patient can learn a few key words, and thereby communicate his or her needs.

Occlusion of the central branch leads to weakness of the face and arm, with minimal non-fluent dysphasia.

Occlusion of the posterior parietal branch produces fluent aphasia, with difficulty in comprehending spoken speech. A mild hemiparesis, and cortical sensory loss with sensory 'inattention' occurs. Sensory 'inattention' is the ignoring of one side on bilateral simultaneous stimulation. Sensory signs may be difficult to elicit in some patients because of problems with communication.

Non-dominant hemisphere lesions Occlusion of the precentral and central branches leads to facial and arm weakness only. Occlusion of the posterior parietal and superior temporal branches leads to dressing apraxia and loss of recognition of faces (prosopagnosia). There is also loss of topographical memories. Dressing apraxia results in the patient sometimes putting his arms through only one sleeve of his shirt, and being unaware that he has done so. Prosopagnosia leaves him unable to recognise the faces of his relatives, but he can recognise them by voice. Loss of topographical memory result in the patient being lost in his own home and not finding his way around his neighbourhood. Cortical

sensory loss and minimal hemiparesis may also occur as a result of vascular occlusion.

Localising lesions in a hemiplegic patient Features that help to localise a cortical, capsular, and middle cerebral artery territory lesions have already been discussed above. If the lesion is in the anterior cerebral artery the weakness is greater in the leg due to involvement of the parasagittal area. Middle cerebral artery lesions leave the arm weaker than the leg. Lesions of the brainstem at the level of the midbrain cause either lethargy, coma and/or impairment of upward gaze. A lesion that involves the third nerve nucleus results in an ipsilateral third nerve palsy and a contralateral hemiparesis - the Weber's syndrome. When the infarct involves the red nucleus, a coarse tremor develops in the arm on the side of the lesion, with a contralateral hemiplegia Benedikt's syndrome.

Lesions in the pons that involve the vestibular nuclei cause vertigo at the onset. The Millard-Gubler syndrome consists of sixth nerve palsy, and facial palsy on the side of the lesion with a contralateral hemiplegia. Foville's syndrome is similar to the Millard-Gubler syndrome, except that a conjugate gaze palsy replaces the sixth nerve palsy. A lesion of the medial medulla causes ipsilateral paralysis of the muscles of the tongue, and a contralateral hemiplegia that spares the face. Loss of postural sensibility also occurs. A lesion of the lateral medulla results in difficulty in swallowing and a Horner's syndrome and cerebellar signs ipsilaterally. Facial sensation is lost on the same side while there is loss of sensation on the opposite side of the body. This syndrome is known as the syndrome of the posterior inferior cerebellar artery, or Wallenberg's syndrome. In lesions of the upper cervical cord a pure motor spastic hemiplegia with sparing of the cranial nerves is seen. The table below summarises clinical findings that occur following lesions of the pyramidal tract at various levels.

TABLE 3.2

NEUROLOGIC DEFICITS RESULTING FROM LESIONS OF
PYRAMIDAL TRACTS AT DIFFERENT SITES.

Cortical lesion	Flaccid hemiplegia with focal convulsions and other cortical signs such as aphasia, apraxia etc.
Internal capsule	Spastic hemiplegia with or without sensory involvement and/or hemianopia.

Upper mid brain	Superior gaze palsy with hemiplegia.
Mid brain	Third nerve with contralateral hemiplegia (Weber's syndrome). Red nucleus tremors with contralateral hemiplegia.
Pons	Vertigo at onset, 6th and/or 7th nerve with contralateral hemiplegia (Millard-Gublar syndrome). Conjugate gaze palsy with contralateral hemiplegia (Foville's syndrome)
Medial medulla	Ipsilateral tongue with contralateral hemiplegia and posterior column involvement.
Lateral medulla	Soft palate, vocal cords, Horner's syndrome, Vth sensory loss, cerebellar signs and contralateral hemianaesthesia (Syndrome of posterior inferior cerebellar artery).
Upper cervical cord	Spastic hemiplegia without cranial nerve involvement.

Aetiology In the vast majority of cases hemiplegia results from a vascular lesion whether of an embolic, thrombotic, or haemorrhagic nature. The lesion can affect either the carotid or basilar circulation. Embolic lesions begin suddenly and may even present as a focal seizure. There may be a history of transient ischaemic attacks. Emboli may arise from the heart as in mitral stenosis, auricular fibrillation, subacute bacterial endocarditis, or following myocardial infarction. Emboli have also been described in atrial myxomas, and mitral valve prolapse. Emboli may also arise from marantic thrombi on the cardiac valves in patients with carcinoma. Common sources of emboli however, are debris from plaques, of atherosclerosis in the large arteries supplying the brain. Atherosclerotic plaques tend to occur at the bifurcations of major arteries, such as carotid bifurcation. Polycythaemia is a rare cause of transient ischaemic attacks. Emboli are said to occur more commonly in the territory of the vertebral artery. Ischaemic lesions that are secondary to atheroma, syphilis, or tubercular arteritis result in thrombosis. Thrombosis comes on gradually, most commonly whilst the patient is asleep. Hypertension, diabetes, obesity, and hyperlipidaemia are the major "risk factors" that increase the likelihood of "stroke". Hemiplegia may also occur as a result of tumours of the central nervous system such as gliomas, oligodendrogliomas, and in metastatic lesions. Arteriovenous malformations, viral encephalitis, and cerebral malaria are rarer causes of stroke. Cerebral venous thrombosis is seen in the postpartum period and in some women who use oral contraceptives.

The prognosis of hemiplegia depends upon the initial deficit and the rate at which function is regained. If the patient presents in a coma the prognosis is likely to be bad. Pinpoint pupils, persistent flaccidity, accompanying aphasia, all adversely affect recovery. Considerable neurologic deficit results from such lesions.

Management of a case of hemiplegia depends upon the causative factor. The nature of the lesion, embolic or haemorrhagic, also decides the line of therapy.

Haemorrhagic infarct Nursing care is most important, airway should be kept clear, the patient turned frequently to prevent bed sores and a nasogastric tube should be inserted to allow feeding.

Cerebral Haemorrhage The commonest cause of cerebral haemorrhage is rupture of a micro aneurysm in a patient with a history of hypertension. It is more common in males than in females. The internal capsule is the commonest site of haemorrhage. Coma develops rapidly, the pulse rate is slow, the breathing is stertorous. 'Cheyne-Stokes' respiration may occur. The head and eyes look at the lesion. If blood has seeped into the subarachnoid space, the patient may be markedly restless. He may move the unaffected side continuously. The hemiplegia is initially flaccid. Cerebral haemorrhage was previously said to have a mortality of 70%, but recent studies which use computerised tomography to classify 'Stroke' show a mortality closer to 45%. This lower mortality is due a better diagnostic capability. Many clinically diagnosed cases of cerebral infarction have been proven to be small haemorrhages on C.T. scan. Pontine haemorrhage results in facial paralysis on the side of the lesion, and hemiplegia on the contralateral side. The eyes look to the opposite side due to loss of conjugate gaze on the side of the lesion. Initially the signs may be unilateral but as the haemorrhage extends the signs become bilateral and extensive. The patient is comatose, with pinpoint pupils, bilateral facial weakness, loss of eye movements, quadriplegia and bilateral extensor plantar responses. No gag reflex is elicited and there is inability to swallow. Haemorrhages generally develop rapidly, when the patient is active during the day. Loss of consciousness occurs early in the course of the disease. If there is blood in the C.S.F. it suggests that there has been extension of the haemorrhage into the subarachnoid space.

Primary subarachnoid haemorrhage is differentiated from intracerebral haemorrhage by the initial symptoms. There is a sudden onset of

severe unrelenting headache, vomiting may also occur. Signs of meningeal irritation such as neck stiffness, Kernig's sign and Brudzinski's sign are present. When cerebral haemorrhage occurs in a younger, normotensive individual a bleed into a tumour should be suspected.

Early coma, pinpoint pupils and extensive bilateral neurologic deficit are bad prognostic signs. An indwelling urinary catheter should be inserted, or condom catheter drainage should be instituted. Hypertension should be effectively controlled by the use of drugs such as Hydralazine. Intravenous diuretic therapy using Frusemide, and in severe cases a continuous drip of sodium nitroprusside may be necessary to lower the blood pressure. It is also necessary to correct any electrolyte and acid-base disturbance that may be present. There is no specific therapy for cerebral haemorrhage. Haemorrhages that are close to the surface and are on the non-dominant side may be removed surgically. Ischaemic lesions should be divided into two categories, the 'stroke in evolution' and the completed stroke. The general management for both these conditions is the same as in cerebral haemorrhage. Anticoagulation has been tried in patients with an evolving stroke. The use of anticoagulants is still a matter of controversy. There is no unequivocal evidence that anticoagulation is of any benefit in either the 'stroke in evolution' or the completed stroke. If the neurologic deficit is minimal, if cerebral haemorrhage can be effectively ruled out, and if there is no obvious contraindication to anticoagulation, a coagulation profile that includes bleeding, clotting, and prothrombin time should be done. Heparin is then given, intravenously, an initial dose of 10,000 units is followed by a continuous drip in which the dose of Heparin is titrated according to the changes in the coagulation profile. A dose of 5000 - 7500 units every 4-6 hours is generally needed. One aims at keeping the clotting time at least twice normal. In India, institution of anticoagulant therapy can only be done when supportive laboratory facilities are available. The physician should also assess whether the patient will co-operate with the close monitoring that long term anticoagulation therapy entails. When the stroke has resulted in a deficit that causes a patient to fall often viz. ataxia anticoagulants should not be used. Warfarin should be used for long term therapy, an initial dose of 20 mgms orally, followed by a suitable maintenance dose of 2.5 to 5 mgms is usually required to allow optimal prothrombin time—prolonged to two and a half times the normal. Anticoagulation therapy should be discontinued after six months as prolonged anticoagulation is associated with a high risk of iatrogenic cerebral haemorrhage. Acetylsalicylic acid has been shown to prevent transient ischaemic attacks in men, a dose of two 300 mgm tablets being taken twice a day.

Simultaneously, measures to reduce cerebral oedema are instituted, 350 ml of intravenous low molecular weight dextran followed by 250 ml of Mannitol is given daily for five days. The efficacy of these measures is as yet unproven. Since many patients with cerebrovascular disease are diabetic and hypertensive, appropriate measures to treat the underlying cause have also to be instituted. Steroids have not proven useful in the treatment of completed stroke, but may be used to relieve life threatening oedema.

No cerebral vasodilators are available that will improve cerebral circulation in the infarcted area. In spite of the major advances in Neurology in the past 25 years, there is little to offer therapeutically to the patient who has a completed stroke. Physiotherapy and rehabilitation measures have much to offer and should be started as soon as possible. Passive movement of the affected limbs should be started on the first day.

PARAPLEGIA Paraplegia can result from disease of muscle, blockage at the neuromuscular junction, or lesions of the lower and upper motor neurone. By common usage however, it has come to denote paralysis of the lower limbs due to a lesion of the spinal cord. The terms spastic paraplegia and flaccid paraplegia are used depending on the tone of the muscles. When incoordination of movement accompanies a spastic paraplegia, the term ataxic spastic paraplegia is used.

CLINICAL FEATURES OF UPPER AND LOWER MOTOR NEURONE PARALYSIS

'Neural shock' at the onset of an upper motor neurone lesion leaves the limbs flaccid and immobile. "Spinal shock" produces loss of reflexes, and severe disturbance of bladder and bowel function. The spinal shock is more pronounced in the humans. The causes of spinal shock are many though the exact pathophysiology of the phenomenon is not understood. As the spinal shock subsides, flaccidity gives way to spasticity, and reflexes become brisk, with patellar and ankle clonus. The plantar response previously unelicitable, becomes clearly extensor. A complete transection of the spinal cord leaves segments below the lesion, free of any higher influences. The flexor tone increases and minimal sensory stimulation results in flexor spasm and evacuation of the bladder and bowels. Paraplegia-in-flexion generally tends to occur after a mid-thoracic transection of the cord. Paraplegia-in extension, which is due to preservation of postural reflexes, occurs after a cervical transection, whilst upper thoracic lesions may result in either flexion or extension states.

The table below differentiates between upper and lower motor neurone lesions with respect to the clinical findings.

TABLE 3.

CLINICAL FEATURES OF UPPER AND LOWER MOTOR NEURONE LESIONS

	Upper	Lower
Tone	Spastic	Flaccid
Spasms	May be present	Absent
Fasciculations	Absent	Present
Atrophy	Absent	Present
Reflexes	Exaggerated	Absent or depressed
Type of paralysis	Limbs involved	Muscles involved.
Reaction of degeneration	Absent	Present

Lower motor neuron lesions result in flaccidity, the muscles undergoing rapid atrophy. The deep tendon reflexes are lost, Fasciculations are marked in anterior horn cell disease, and can be seen as contractions of muscle fibre bundles. These movements do not cross a joint and are thus differentiated from myoclonus. The movements has been described as similar to the movement seen in 'a bag of worms'. Spontaneous contractions of individual muscle fibres are not visible to the naked eye except in the tongue and are know as fibrillations. They are best studied by electromyography.

Weakness due to lower motor neurone disease may sometimes be difficult to differentiate from that due to primary muscle disease. Myopathy denotes primary disease of muscle, without involvement of the central or peripheral nervous system. Weakness due to disease at the neuromuscular junction results in 'myasthenia gravis' which can rarely be confused with lower motor neurone disease. Primary disorders of muscle generally lead to selective involvement of proximal musculature, and are associated with atrophy. Electromyography can be a great help in the diagnosis of these conditions.

Symptoms such as ataxia, loss of sensation and sphincter disturbances, are often associated with paraplegia, depending on the cause

and the site of the lesion. Ataxia, in spinal cord disease, may be due to involvement of spinocerebellar fibres, or the posterior columns, in which case the ataxia is designated as 'sensory ataxia'. Spinothalamic tract involvement results in loss of pain and temperature sensation a few segments below the level of the lesion. Paralysis of respiratory muscle function occurs in high cervical lesions.

There is loss of autonomic function in the phase of 'spinal shock', which increase the chance of a bed sore. The bladder is totally paralysed during the phase of 'spinal shock', leading to urinary retention. As the shock subsides, the bladder starts to contract reflexly and an "automatic bladder" is established. Sexual function is also impaired, higher psychic stimuli failing to excite the sexual organs. However, stimulation of the sexual organs themselves may lead to reflex penile erection. Sphincter disturbance can lead to a total retention of urine, or to hesitancy, where the patient has to strain to pass urine, or there may be reflex voiding.

ANATOMY AND PHYSIOLOGY OF MICTURITION Sphincters control the flow of urine from the bladder to the urethra. The internal sphincter is not a true sphincter in both sexes. It is the arrangement of smooth muscle fibres, and elastic tissues that keep the urethra in a collapsed state. The angle that the bladder makes with the urethra also helps to keep the urethra closed. The external sphincter, on the other hand, is a true sphincter, but, it is not strong enough to keep the patient continent. It is located at the level of the uro-genital diaphragm, and is under voluntary control.

Parasympathetic nerves from the sacral segments of the cord (S 2, 3) provide the motor supply to the detrusor muscle of the bladder. These fibres travel with the internal pudendal nerve. The sympathetic supply comes via the hypogastric plexus. The exact nature of sympathetic control of bladder is not understood but is thought to affect the external sphincter.

Afferent fibres from the stretch receptors form a reflex arc with the centre in the third sacral segment. These fibres also travel in the posterior columns to the cortical centre for inhibition located in the frontal lobe. Fibres descend from this centre, lying adjacent to the pyramidal fibres. The stretch receptors are stimulated according to the quantity and weight of urine in the bladder. This explains the desire to pass urine on arising in the morning, as the afferents are more active in the upright position than when lying down. The sacral reflex arc

controls micturition in infancy, senility and in the unconscious state. Cortical centres exert an inhibitory influence over this reflex mechanism, through voluntary control. This influence also affects the hypothalamic and pontine facilitatory centres, and the muscles of the floor of the pelvis.

Micturition involves relaxation of cortical inhibitory control, with the relaxation of the external sphincter and pelvic floor. The sacral reflex now takes over, the detrusor muscle contracts, pulling open the bladder neck, the internal sphincter relaxes, and urine flows out due to a rise in intravesical pressure, and due to the effects of gravity.

PRINCIPAL TYPES OF NEUROGENIC BLADDER

Uninhibited bladder It results from loss of cortical inhibitory control. A tumour of the paracentral lobule, Alzheimer's disease, and superior sagittal sinus thrombosis are some causes that lead to an uninhibited bladder. There may be urgency and hesitancy, with the patient voiding precipitously. Bladder sensation is normal, and there is no residual urine. The capacity of the bladder is moderately reduced.

The spinal or automatic bladder It results from disease that affect the spinal cord above the sacral segments. The bladder is small, sensation is vague and poorly localised. The patient voids suddenly and reflexly. There is a small amount of residual urine after voiding. Some awareness of bladder evacuation is felt. When a flexor spasm gives rise to a 'mass reflex' there may be involuntary evacuation of bladder and bowels, associated with autonomic disturbances such as sweating, and pallor.

The autonomous bladder It is a bladder without any external innervation. It is caused by lesions of the sacral spinal cord, conus medullaris, and cauda equina. There is destruction of the parasympathetic nerve supply, with absence of bladder sensation. No reflex or voluntary control is present, the bladder empties incompletely with continuous dribbling of urine, and a large amount of residual urine. Bladder capacity is however not markedly increased.

The sensory bladder It is seen in tabes dorsalis, and rarely, in diabetes mellitus. There is no bladder sensation and the capacity of the bladder is markedly increased. The patient voids by straining, there is overflow incontinence, with a large amount of residual urine. This bladder is also known as an atonic bladder.

The table below lists some causes of paraplegia and correlates them with sites in the neuraxis.

TABLE 3.4

CAUSES OF PARAPLEGIA

- A) Intra cranial causes :**
- 1) Cerebral diplegia
 - 2) Hydrocephalus
 - 3) Wounds of motor cortex in the parasagittal region.
 - 4) Thrombosis of the superior sagittal sinus
 - 5) General paralysis of the insane.
 - 6) Tumours of the falx.
- B) Spinal causes**
- Primary cord disease.
- 1) Subacute combined degeneration.
 - 2) Disseminated sclerosis.
 - 3) Motor neurone disease.
 - 4) Syringomyelia
 - 5) Hereditary ataxias.
- Secondary affection of the cord
- a) Trauma
- 1) Fracture dislocation of vertebra
 - 2) Congenital dislocation of atlas
 - 3) Penetrating wounds.
 - 4) Concussion of the cord
 - 5) Electric shocks and lightning.
- b) Infection
- 1) Viral transverse myelitis
 - 2) Syphilitic meningo-myelitis
 - 3) Tuberculous arachnoiditis and other tuberculous affections tuberculoma.
 - 4) Post infectious myelitis like measles chicken pox.
 - 5) Post vaccinia, serum.
- c) Vascular
- 1) Haemorrhage i) Intra thecal (spinal epidural haemorrhage)
ii) Intra-medullary (Haematomyelia)
iii) AV malformations.
 - 2) Thrombosis (Anterior spinal artery thrombosis).
 - 3) Embolic (Very rare).

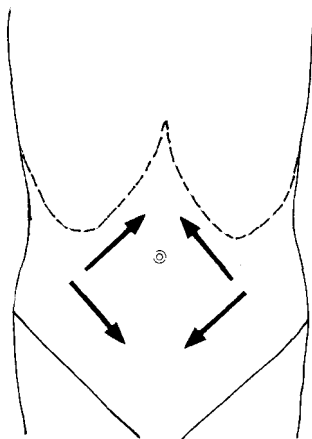
The differential diagnosis of weakness and wasting of the small muscles of the hands may at times be problematic. This may occur in

multiple conditions with varying levels of lesion, and can result from diseases of muscle, peripheral nerves, radicles or lesions of the anterior horn cells.

Weakness due to distal dystrophy is symmetrical in both hands without any sensory loss. Proximal musculature may also be involved. With lesions of the peripheral nerves, the commonest in our country being leprosy, there is associated sensory loss and trophic changes in the distribution of the nerve, in this case the ulnar nerve. Lead neuropathy causes unilateral weakness with a wrist drop. Hereditary sensory motor neuropathy of the Charcot Marie Tooth type initially involves the lower limbs but may involve the upper limbs. The wasting extends to the lower third or middle of the forearm. Pain is an important feature in brachial plexitis or thoracic outlet syndrome. Addson's test (described later) will be positive, in the latter condition. Patients may be left with wasting and weakness of the hands following a severe attack of Guillaine-Barre type of infective polyneuritis. History will make the diagnosis obvious. Syphilitic and tuberculous arachnoiditis can be recognised by the associated cord involvement and CSF examination. Anterior horn cell lesion occurs in motor neurone disease, poliomyelitis syringomyelia and crano-vertebral anomaly. Presence of fasciculations in other musculature help in the diagnosis of motor neurone disease. Poliomyelitis rarely ever affects the small muscles of the hand. However, it begins as an acute illness with fever and the affection is asymmetrical. Syringomyelia has loss of reflexes in the upper limbs, brisk reflexes in the lower limbs and classical dissociated and hanging anaesthesia. Short neck, low hair line, torticollis and ataxia with posterior column signs are the associated findings in crano-vertebral anomalies.

REFLEXES

Reflexes are of two types, the superficial and the deep. Superficial reflexes include the corneal, abdominal, cremasteric, and anal reflexes. The cremasteric and anal reflexes are elicited when a lesion of the spinal cord is suspected. The corneal reflex has been already described under the section on cranial nerves. The abdominal reflex should be elicited with a sharp object, with care being taken that no blood is drawn as a result of the test. Moderate pressure is used, and the examiner observes the direction of movement of the umbilicus as each quadrant is tested. The movement of the umbilicus is shown by the direction of the arrows in the Figure 3.2.



Obtaining deep tendon reflexes is an art that needs to be mastered when the student is doing his clinical rotations. The novice has to learn to use the reflex hammer. It must not be held tightly as this prevents the swing action of the hammer. The handle of the hammer should be held with the thumb and index finger, the fist lightly closed over the arm of the hammer. The tendon is tapped and not hit. It is important that the tendon have a certain stretch applied to it. When the ankle jerk is elicited the foot is lightly held in one hand, the patient is asked to relax, the tendoachiles is stretched slightly and then tapped with the hammer. The knee jerk is best elicited by asking the patient to relax, and flex his knee. The examiner then taps the quadriceps tendon briskly. The biceps jerk is tested by asking the patient to flex his arm at the elbow, and the examiner taps the tendon, placing his finger over the tendon. The supinator jerk is elicited by holding the patient's forearm semipronated in the left hand and striking the hammer over the distal end of the radius. The triceps jerk is elicited by flexing the elbow a little over 90 degrees, to stretch the triceps tendon.

Deep tendon reflexes are monosynaptic reflexes which are affected in lesions of either the afferent or the efferent arc. They are inhibited by the pyramidal system, therefore in lesions of this system they become exaggerated. The segments controlling the reflexes are: C5, 6 for the biceps and the supinator, C 5,6,7, for the triceps, L 2,3,4, for the knee, and L 5,S 1, for the ankle reflexes.

Exaggeration of reflex activity may lead to an overflow resulting in contraction of muscles supplied by adjacent spinal segments. An inversion of the reflex is said to occur when the reflex is absent in the appropriate segment, but an overflow is still present in adjacently supplied muscles.

The jaw jerk is elicited by asking the patient to open his mouth. The examiner places his index finger of his hand on the patient's chin and strikes the hammer over the finger. The jaw jerk is exaggerated in upper motor neurone lesions above the level of the pons.

The plantar response should be elicited with the use of a rather pointed object like the end of a car key, or the blunt end of a reflex hammer. An object sharp enough to draw the patient's blood in the elicitation of the response is *Not* to be used. Stroke the lateral border of the foot move upwards and medially towards the ball of the foot. Make sure the patient is aware of what you are about to do, and is relaxed then you elicit the response.

A reflex response should be characterised as absent, normal, hypoactive, or exaggerated. Bilaterally symmetrical brisk reflexes generally are of no clinical significance. When the reflexes are asymmetrical, decide which is the abnormal side, and report the response in terms of the affected side, e.g. the reflexes are hypoactive on the left. It may sometimes be difficult to decide the side of the abnormality on the basis of the reflexes alone. Additional features in the history and the physical examination will help in localising the abnormality.

CHAPTER IV

CEREBELLAR AND SENSORY SYSTEMS

COORDINATION

Tests for coordination include the finger to nose and heel to shin tests. In the finger to nose test, the patient is asked to touch the tip of his/her nose with the tip of his/her index finger, and then to touch the examiner's finger which is held in front of his/her at a distance of half a metre. The position of the examiner's finger after each touch is changed in both horizontal and vertical axis. If the patient has a field defect, the test must be conducted in the normal field. Observe whether the target is approached accurately, or if tremor develops on approaching the target. Missing the target is known as 'pastpointing', whilst the tremor which develops as the target is approached is called 'intention tremor'. For the test of posterior column function, the patient is requested to close the eyes, his/her index finger is placed in space with his/her limb fully extended, and then asked to touch his/her nose. With posterior column lesion, the patient gropes about on reaching the target or misses the target. The heel to shin test involves tapping one knee with the other heel and then sliding the heel down the shin smoothly and accurately, with eyes closed. The first part of the test assesses the posterior column function and the heel-shin part evaluates the cerebellar function.

Other tests of coordination include rapid alternating movement of the hands by pronation and supination at the elbow and tandem gait. Tandem gait is tested by asking the patient to walk on a straight line with one foot kept directly in front of the other. Inability to walk in a straight line with one foot directly in front of the other; is often the first sign of cerebellar disease. In cerebellar lesions, the patient stands on a wide base and on reducing the base by bringing two feet together, he/she will sway or fall despite the eyes being open. In posterior column lesion the patient is unable to stand steady with two feet together with eyes closed. This is designated as Romberg's sign.

Intention tremor and pastpointing, as well as difficulty with the heel to shin test must be judged in terms of differences between the two sides. The affected side points to an ipsilateral cerebellar disturbance. Minimal muscle weakness can be confused with cerebellar disease and

careful examination alone will differentiate the two to the newcomer. Trunkal ataxia is instability of the trunk, the patient being unable to sit or stand without swaying. The cerebellum is divided into a midline vermis and two lateral hemispheres. A dictum, that the student must remember, is that ataxia is a sign of midline cerebellar disease, whilst disturbance in finger to nose movement implies disease of the cerebellar hemispheres. Also remember that hemispheric disease may show midline signs but never vice versa. Other signs of cerebellar disease are hypotonia, nystagmus, and staccato scanning speech. Cerebellar signs are also found in lesions that involve the connections of the cerebellum, within the brainstem. Trunkal ataxia is usually due to a lesion of the vermis and its connections. It is seen with medulloblastomas, a malignant tumour, common in childhood.

DISEASES OF THE CEREBELLUM

Cerebellar diseases in childhood are very different from those seen in adults.

Cerebellar maldevelopment It is often associated with the Dandy-Walker syndrome and the Arnold-Chiari malformation. In the Dandy-Walker syndrome the exit foramina of the ventricles, namely the foramen of Luschka and Magendie, that develop at the seventh week of gestation, fail to open. It results in dilatation of the fourth ventricle, which enlarges, and leaves no space for the growing cerebellum. A massive midline cyst, the original fourth ventricle, develops instead, with atrophic cerebellar tissue, lying beside the cyst. Despite the extent of the maldevelopment, there may be no symptoms till as late as the second decade of life. Signs of cerebellar lesions in infancy become apparent around two years of age, as the pyramidal pathways become functional. The Dandy-Walker syndrome usually presents with signs of obstructive hydrocephalus, such as an enlarging head, headaches, vomiting and papilloedema.

The Arnold-Chiari malformation may be a result of pontine maldevelopment. The pontine flexure does not form, and a part of the cerebellum and medulla, is pushed down into the cervical canal. The malformation becomes symptomatic at any time, from two years till late in the third decade of life. It may be present with signs of an obstructive hydrocephalus, or with a combination of signs such as lower cranial nerve palsies, ataxia, spastic quadriparesis, and deficits in proprioception involving the upper extremities. Abnormalities of the

cranio-vertebral region, namely basilar invagination, occipitalisation of the atlas, maldevelopment of the atlas with atlanto-axial dislocation, etc. are often seen in our country. These conditions should always be remembered when one is examining a patient with cerebellar disease. A short neck and a low hairline are often observed with these conditions. Basilar invagination is associated with a small posterior fossa, and herniation of the cerebellar tonsils into the upper cervical canal.

Metabolic disorders Many of the cerebellar diseases of childhood present as acute attacks of ataxia, that recur, and where no cause can be found. The child is free of symptoms after two or three attacks, which generally occur after the first year of life and continue throughout the first decade of life. An individual attack may last for a few days and recur within months or after several years. Maple syrup urine disease, a disorder where increased amounts of Valine, Leucine, and Isoleucine are excreted in the urine, is a fatal disease of childhood that may show cerebellar signs. Hartnup's disease, a disorder of tryptophan metabolism, has cerebellar signs, pellagra like skin, and peripheral neuropathy. Leigh's encephalopathy, a disorder of pyruvate metabolism, is another cause of acute ataxia.

Ataxia telangiectasia is a progressive disorder that affects the cerebellum, in the first or second decade of life. Telangiectasis of the conjunctiva and a tendency to recurrent infections due to agammaglobulinemia are other notable features of this disease. The patient dies as a result of the intercurrent infections.

Ataxia sometimes follows acute viral infections such as chicken-pox, measles, and infectious mononucleosis. It may follow within three to six weeks of infection and continue for a few months. Proximal girdle weakness in children due to pyramidal lesions or nerve root involvement may be confused with ataxia. Friedreich's ataxia, a heredo-degenerative condition always manifests itself in childhood. It is rare in India, variety of olivo-ponto-cerebellar atrophy with slow eye movements is commoner in our country. The hereditary ataxias are classified into three divisions: A) the pure spinal and spino-cerebellar, B) pure cerebellar, C) those associated with peripheral neuropathy.

Pure spinal form Sanger-Brown variety of ataxia is an example of pure spinal ataxia. It is occasionally seen in India, the ataxia involving all four limbs, with speech difficulties occurring later in the course of the disease. Signs indicating pyramidal tract involvement such as hyperre-

flexia, and spasticity are always present. A few cases have optic atrophy, pupillary changes, and oculomotor palsies.

Spino-cerebellar ataxia Friedreich's ataxia is a spino-cerebellar form of ataxia with autosomal recessive inheritance. The symptoms manifest themselves at the age of 8-10 years. It usually begins as a progressive gait disturbance with the ataxia involving the upper limbs after a few years. Later speech is also affected. On examination the child may have optic atrophy, nystagmus, staccato speech, ataxia of all four limbs, brisk deep tendon reflexes, except for absent ankle jerks, and extensor plantar responses. There is loss of proprioception in the toes. Kyphoscoliosis, pes cavus, and hammer toes are present and cardiac abnormalities occur. The electrocardiogram shows prolongation of the P-R interval, bundle branch block, and ST segment changes. Myoclonic and generalised tonic-clonic seizures are known to occur in these patients.

Olive-ponto-cerebellar atrophy with slow eye movements is a spino-cerebellar ataxia of recessive inheritance, that is often seen in India. The condition begins to cause symptoms in the early teenage years, with a progressive gait ataxia. Later the upper limbs and speech becomes involved. There may be mild mental retardation, and bilateral ptosis. Pursuit eye movements are slow. On asking the patient to look to the right or left, he initiates the movement with a jerk of the head, the eyes following subsequently. Optic atrophy is rare, whilst signs of pyramidal involvement are usually present. Proprioceptive deficits, peripheral neuropathy, and extra-pyramidal signs may be present in varying combinations. Kyphoscoliosis is present, but pes cavus is usually not seen.

Pure cerebellar forms These occur at any age. The Ramsay-Hunt syndrome, which begins in the third decade of life, is associated with ataxia, intention myoclonus and epilepsy. When there is mental retardation and progressive deterioration of gait, Lafora body disease may be suspected. Primary cerebellar degeneration is seen in the fifth and sixth decades of life. Cerebellar disease can be a non-metastatic presentation of several malignancies, notably carcinoma of the lung, breast, and prostate. In India, rarely, ataxia can be due to salmonella infection.

Diphenylhydantoin (DPH) is known to cause cerebellar degeneration, after chronic use. However, it is not certain whether the degeneration is secondary to frequent seizures or to chronic drug use. Acute DPH toxicity leads to a reversible form of ataxia. Alcohol is a major cause of

cerebellar ataxia and tends to affect the vermis only. The patient has ataxic gait and even trunkal ataxia but no disturbance of coordination involving the upper limbs. Prolonged fasts and starvation are other causes of cerebellar degeneration. The cerebellum is a common site of tuberculomas, and of primary and metastatic neoplasms.

Cerebellar disease in adults Ataxia of acute onset in the adult usually means a haemorrhage into the cerebellar parenchyma, either primary or secondary to an arterio-venous malformation. A severe occipital headache, vomiting and ataxia herald the onset of this catastrophic illness. Papilloedema and deterioration in consciousness occur rapidly due to the development of an acute obstructive hydrocephalus. Death may occur if the blood clot is not evacuated immediately. The diagnosis is established by angiography, or on computerised tomography.

Ataxia due to degenerative disease is usually of the olivo-ponto-cerebellar type in India, with Marie's and Sanger-Brown's variety being next most common. Marie's ataxia has posterior column dysfunction in addition to cerebellar signs. Sanger-Brown's ataxia has been described in the earlier section.

Multiple sclerosis is an important and common cause of cerebellar dysfunction. It is associated with visual loss, brainstem and spinal cord involvement. It typically presents in the late twenties as a unilateral loss of vision. The patient then has a relapsing and remitting course with spinal cord and cerebellar signs developing as the disease progresses. The initial loss of vision is rapid, with the vision reduced to mere perception of light, or even total blindness, in a matter of hours. This disease is called retrobulbar or optic neuritis, and generally recovers almost completely. The ophthalmoscopic examination is initially normal, though later optic atrophy and a Marcus-Gunn pupil are seen. The patient complains of pain on moving the eyes. Spinal cord and brainstem dysfunction cause pyramidal signs, sphincter disturbances, and diplopia or dysphagia. There are some cases with multiple sclerosis that present with a complete transection of the cord, and others that show only brainstem involvement.

Over many years the unfortunate patient is left with substantial neurologic deficit and is crippled by the disease. Death is generally due to intercurrent infection. The diagnosis is firmly established if there is a history of remissions and relapses, and signs point to lesions in the neuraxis at various levels. CSF examination shows an increase in gamma

globulin, and presence of myelin basic protein. Visual auditory and somato-sensory evoked potentials are also abnormal in the vast majority of patients.

The aetiology of this cruel illness is unknown though some evidence point to a viral source. Multiple sclerosis is a disease of temperate climates and affects peoples who live in the northern hemisphere. It also seems to affect individuals from 'high risk' areas who migrate to 'low risk' areas after the age of fourteen. A rise in measles titres has been demonstrated in patients with multiple sclerosis. However, the viral aetiology is as yet unproven.

There is no specific treatment for this disease but steroids may shorten the relapse and speed recovery. Symptomatic measures including the use of antispasmodic agents such as Baclofen help to relieve the patient's distress to some extent. Physical therapy and occupational therapy should also be utilised.

Devic's disease is a demyelinating disease that is seen more commonly in India, than multiple sclerosis. It occurs in a younger age group, 10-20 years of age, and has a predilection for males. Retrobulbar neuritis affecting one or both eyes precedes, or follows, the onset of transverse myelitis that affects the cervical or thoracic spinal cord. The patient may recover from the visual loss and the paraplegia within a period of two to three weeks. A few patients, however, are left with visual deficit and paralysis.

DISORDERS OF GAIT Normal gait consists of coordinated painless movements of the pelvis and lower extremities, associated with rhythmical movements of the arms, trunk and head. Gait, therefore, can be abnormal in disorders that affect coordination, tone, power, and in painful lesions of joints and muscles. We shall now describe specific gait disorders.

Cerebellar gait This gait resembles that of a drunken men. The patient reels and sways from side to side, keeping his legs wide apart to broaden his base. There is a tendency to fall to the side of the lesion; if only one lobe of the cerebellum is involved in the disease process, the patient cannot stand on the homolateral foot. The degree of instability varies with the extent and duration of the disease. Sometimes there is only mild swaying that is best observed when the patient turns, whilst in others the disease leaves the patient with severe ataxia so that he is

unable to sit up. Blindfolding the patient does not aggravate his gait disturbance.

Tabetic gait is seen in lesions of the posterior columns. A mild case of *tabes dorsalis* complains of problems with gait only in the dark, because he is otherwise able to compensate visually for the proprioceptive loss. Blindfolding the patient makes him ataxic, there being no visual compensation to maintain balance. A severe case of *tabes* stands on a wide base and guides his feet by visual means. The absence of proprioceptive input leaves the higher centres totally dependant on the eyes for spatial information. The classical 'high steppage gait' of *tabes* is seen very late in the disease. The patient is unable to walk even short distances without support. The foot is raised far off the ground and then brought down with a thump because the patient misjudges the distance between the ground and his feet.

Peripheral neuritis Here both afferent and efferent pathways are involved resulting in a gait that has to compensate for ataxia and weakness. The feet barely clear the ground due to foot drop. The toes scrape the ground as the feet advance and the patient walks with a wide base. Flat feet result from the weakness of the strap muscles. The gait is much worse on eye closure.

Hemiplegic gait is known as the circumducting gait, because the paralysed lower extremity makes an outer semicircle as it progresses forwards due to an inability to flex at the knee. Remember that extensor tone dominates in the lower limb in hemiplegia. The paralysed arm does not swing normally, but remains adducted at the shoulder and flexed at the elbow, with a wrist drop. In cases of infantile hemiplegia the patient has greater unsteadiness of gait because of shortening of the limb.

Gait in double hemiplegia, diplegia, or spastic paraplegia The patient crosses her legs due to adductor spasticity of the lower limbs. The adduction at the hips may lead the legs to rub each other or be actually crossed. The gait resembles the movements of the two arms of a pair of scissors, hence the name "scissor gait". When the feet remain plantar flexed the patient walks on her toes. In India lathyrism is often a cause for this gait.

The gait of Parkinson's disease The patient walks with a shuffling, hesitant or festinant gait. The steps are taken slowly and many small steps are taken as though the patient were unsure of himself. The gait is

devoid of springiness and suppleness. The head is held stiffly, the body stooped forwards and the patient shuffles along slowly, displacing his centre of gravity as little as possible. If his foot gets caught in something as he walks, he may fall if he is unable to recover his centre of gravity. The hurried movement forwards to recover the lost centre of gravity is called 'propulsion', whilst a backward movement is called 'retropulsion'.

Myopathic or waddling gait Patients with myopathy who have marked weakness of the pelvic musculature, have difficulty in getting up from the squatting position. They also have problems going up a flight of stairs. They find it extremely difficult to jump and skip. The patient stands with a wide base, and has an increase in the lumbar lordosis so that the upper part of the trunk is thrown backwards. Weakness of the hip flexors results in inability to clear the ground with the feet. The pelvis on the side of the advancing foot is raised by tilting the body to the opposite side. This side to side movement that enables the body to advance forwards is described as 'waddling'.

Hysterical gait It is a very bizarre type of gait, that does not resemble any of the specific gait disturbances described above. The efforts of a hysterical individual are directed towards increasing the abnormality of gait whilst walking, to the extent of falling continuously. A person with a gait disturbance due to an 'organic' condition makes a sincere effort to walk and maintain her balance. A hysterical patient, is unable to take even a few steps with support and gives up easily. He falls often but the falls result in no serious injury. The abnormality increases when the patient is aware that he is being observed.

Gait disturbances are also seen in a variety of basal ganglia disorders such as torsion dystonia, chorea, and athetosis. These gaits have no specific characteristics and differ from patient to patient, depending upon the severity of the disease and the disability that it produces.

SENSORY SYSTEM

Fibres carrying different modalities of sensation reach the root entry zone of the spinal cord via the posterior root ganglia.

The sensation of touch is carried by two types of finely myelinated nerve fibres. Those fibres that carry accurately localised touch travel via the posterior columns, whilst those carrying crude touch cross over to

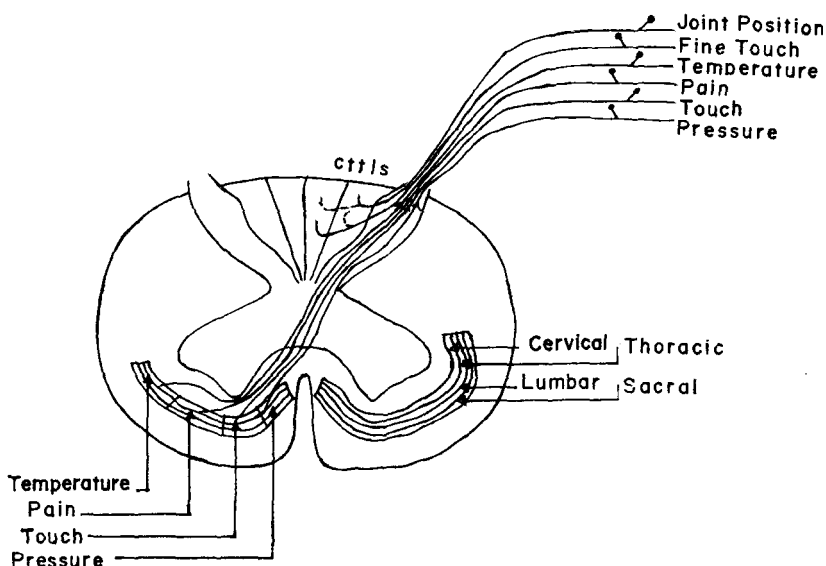


Fig. 4.1 : Diagram to illustrate the arrangement of sensory fibres in the spinal cord.

the opposite side and ascend upwards in the ventral spino-thalamic tract. Fibres carrying pain and temperature also cross in the anterior commissure and travel in the dorsal spino-thalamic tract. The dorsal spino-thalamic tract is arranged in a segmental manner so that fibres from the lower extremities are pushed outwards by newly arriving fibres. The sacral fibres therefore come to lie most laterally, and the cervical fibres medially, in the cervical spinal cord. This segmental arrangement of fibres has clinical significance. An intramedullary tumour of the cord is likely to spare sensation in the sacral area until it has grown to a very large size. The spino-thalamic tract ascends to the brainstem where it is joined by the medial lemniscus carrying the proprioceptive fibres.

Fibres carrying joint and position sense travel ipsilaterally in the posterior columns of the spinal cord. The medially located carry fibres from the legs while those located laterally carry proprioception from the upper extremities. The posterior columns ascend to the gracile (leg) and cuneate (arm) nuclei of the medulla. Second order fibres decussate,

form the internal arcuate fibres and ascend in the medial lemniscus. They join the spino-thalamic tract and end in the thalamus. Third order fibres from the thalamus reach the primary sensory cortex in the parietal lobe.

TEST OF SENSORY FUNCTION

The sensation of touch is tested by a wisp of cotton wool. Pain is tested with a pin, but *not* with anything that is so sharp that it draws blood by piercing the skin. Temperature sensation is ideally tested by using coned metallic tubes. One tube should contain boiling water, the other crushed ice. The temperature difference in the two tubes is kept very wide so that the patient is able to appreciate the difference easily. However, in clinical practice, if pain is normal, temperature is hardly ever abnormal.

Always begin testing for sensation distally and proceed proximally. Test in the normal areas first, and map out the area of sensory loss and match it with the distribution of a root or nerve. If all 3 modalities of sensation are affected (pain, touch, and temperature) it is more than likely that the disturbance is in the peripheral nerve and not in the spinal cord. In peripheral neuropathy the typical sensory loss is in the so called "glove and stocking" distribution, that is, it is in the hands and feet. The student may find some of these descriptive terms peculiar in their usage. Remember that the terminology evolved in the centres of neurology in Europe, and hence phrases common to that continent are often used to describe symptoms and signs in neurology.

Lesions of the spinal cord can result in a variety of patterns of sensory loss. Syringomyelia leads to a "half cape" pattern of "dissociate" sensory loss with loss of pain and temperature but with touch preserved. Hemisection of the cord or Brown-Sequard syndrome leads to ipsilateral loss of vibratory and position sense, with contralateral loss of pain and temperature. As the spino thalamic tracts cross over whilst ascending for two or three segments, a lesion in the cord produces a sensory loss two to three segments below the level of the lesion. Do not confuse the spinal cord level for the level of the vertebrae. The cord ascends after birth, till in the adult it ends at the level of the first lumbar vertebra. To get corresponding vertebral and spinal cord levels add one to upper thoracic vertebrae, and three to the lower six thoracic vertebrae. Therefore a lesion at the level of the *seventh* thoracic vertebrae will involve the *tenth* thoracic spinal cord segment.

It is very important that the patient cooperates fully with sensory examination. Try and get the patient to relax and if necessary divide the examination into more than one session. It is not necessary to ask the patient to close his eyes during sensory testing as cooperation is better with the eyes open (ADD).

Joint and position sense is tested at the distal interphalangeal joints. The finger or toe is held steady at the metacarpophalangeal joint and the finger or toe is then moved at the interphalangeal joint, by holding it at the side, and not at the nail, to avoid push-pull sensation. If there is an abnormality at the distal joints, examination should proceed proximally till a level of a normal perception of joint sense is reached.

Vibration sense is tested by holding a vibrating tuning fork of 128 frequency at a distal bony point such as the styloid process of the radius or the lateral malleolus. If the vibrations are not felt distally, proceed proximally as with joint position testing. Bony points generally used to test vibration sense include the tibial tubercle, the anterior superior iliac spine, the ribs and clavicles.

SYRINGOMYELIA The word syrinx means a rock-cut tunnel and meylon of course means spinal cord. Syringomyelia then, is a cavity in the spinal cord, that may be congenital or acquired. According to one theory the 'congenital' variety may form secondary to a dynamic obstruction of the exit foramina of the fourth ventricle during foetal development. It leads to a mild degree of prolapse of the cerebellar tonsils which then act like a ball-valve mechanism increasing the pressure in the fourth ventricle. This increase in pressure is transmitted to the central canal of the spinal cord, which eventually expands in the cervical region, leading to a cavity filled with fluid - the syrinx. When the syrinx extends upwards into the medulla, it is called 'syringobulbia'. The acquired variety includes post traumatic, and tumour related syrinx.

The syrinx increases in size very gradually, leading to symptoms in the teenage years or even in the late twenties. There is no predilection for either sex. Pain along the clavicle, and shoulder which is dull aching in character may be the first symptoms. At this stage a careful sensory examination may reveal loss of pain and temperature along the second to fourth thoracic segments. The patient may have noticed this area of sensory loss whilst bathing. The syrinx expand and, destroys the spinal cord, with the fibres of the anterior commissure being affected at an early stage. The deep tendon reflexes are absent in those regions where

the anterior horn cells are affected. Typically the biceps and supinator reflexes are absent, with an 'inverted' radial reflex. The sensory loss spreads to involve the 'half cape' area—an area that involves the shoulder, arms and chest on one side. The sensory loss is of the 'dissociate' variety, with loss of pain and temperature and preservation of touch. The patient may suffer burns to the hands as a result of the anaesthesia. There is wasting of the small muscles of the hand, with enlargement of the cavity in the lower cervical region. The syrinx now acts as a space occupying lesion, producing pressure effects on the descending tracts. A spastic paraparesis, with hyperreflexia and extensor plantar responses is seen at this stage of the disease. The descending sympathetic pathway may also be affected giving rise to a "Horner's syndrome". Sometimes the syndrome is present bilaterally giving rise to difficulty in the diagnosis of the syndrome. When the cavity expands still further, sensory loss extends to involve the thighs, but spares the sacral area, as fibres from the sacral region are the most laterally located in the cord. The clinical manifestations of a syrinx will be better understood if the reader turns to the Fig. 4.1 on page 87 and imagines the effects of a gradually expanding lesion on various structures. The diagnosis may be suspected when the sagittal diameter of the cervical spinal canal is more than 22 mms. Contrast myelography may show tonsillar herniation if films are taken with the patient in the supine position. Air encephalography will show the presence of a mild communicating hydrocephalus, and simultaneous air myelography will show progressive collapse of the cord, on gradual removal of CSF.

The differential diagnosis includes basilar invagination and the Arnold-Chiari malformation. These two conditions act as extramedullary lesions and produce weakness that involves one arm and then proceeds to affect other limbs in a clock wise manner. Sensory loss first affects the lower extremities and not the trunk as in syringomyelia. Posterior column signs are found early in these conditions, and there is generalised hyperreflexia with no loss of reflexes in the upper extremity. Haematomyelia may mimic syringomyelia, but the onset in haematomyelia is dramatically different from syringomyelia. Haematomyelia follows a rupture of an arterio-venous malformation or angioma of the cord, and presents as an acute catastrophic events. The patient is in great pain initially, and may be totally paraplegic in a few hours. Intramedullary tumours can be difficult to differentiate from syringomyelia, but their course is not so gradual and there is no cerebellar tonsillar herniation to demonstrate on myelography.

The treatment of syringomyelia remains controversial. Treatment has to be individualised, and posterior fossa decompression, with, or without previous ventricular shunting has been reported as beneficial in several reports. Drainage of the syrinx cavity in the subarachnoid space is carried out with insertion of the silastic tube or sectioning of the filum terminale is done so as to allow the drainage of the cavity via the central canal to the lower subarachnoid space. However, complications abound and a patient with syringomyelia should always be referred to a centre with reach experience in handling this disease.

SUBACUTE COMBINED DEGENERATION The disease is subacute in onset and there is combined degeneration of two systems, namely the posterior columns and the pyramidal tracts, in the spinal cord.

The condition is caused by deficiency of vitamin B12 due to lack of intrinsic factor with achlorhydria (pernicious anaemia). Vitamin B12 deficiency may also occur due to dietary deficiency (common in India), or its malabsorption due to coeliac disease, tropical sprue, intestinal tuberculosis, regional ileitis, diverticulosis or total or partial gastrectomy.

There is demyelination of the posterior and lateral columns of the spinal cord. The disease may also produce demyelination of the posterior roots and peripheral nerves. The process starts in the dorsal region of the spinal cord, spreading upwards and downwards as the disease progresses.

Since the posterior columns are first involved, the initial symptoms are paraesthesias in the form of tingling and numbness, or pins and needles in the lower limbs. The patient may complain of heaviness or swelling of the distal extremities or he may complain that he is walking on soft ground made of cotton wool. Examination at this stage may reveal minimal diminution in appreciation of joint position at the toes, loss or diminution of vibration sense distally and finally difficulty in discriminating two points. Romberg's sign may be positive. As the disease progresses, sensory ataxia develops due to which the patient may not be able to assume an upright posture or walk in the dark or with eyes closed. If posterior roots or peripheral nerves are involved the patient may not be able to appreciate light touch distally (stocking distribution), may have tenderness of calves and loss of ankle jerks.

Lateral column involvement leads to spastic weakness of the lower limbs. The abdominal and cremastic reflexes are lost, the knee jerks are brisk with loss of ankle jerks and extensor plantar responses.

Investigations reveal histamine fast achlorhydria and macrocytic anaemia with a megaloblastic bone marrow. Serum vitamin B12 levels are below 100-200 pg/ml. Schilling test may confirm the diminished absorption of vitamin B12.

If treatment is started early in the course of the disease, full neurological recovery ensues. Vitamin B12 is administered parentally, in the dose of 100 microgramme, thrice weekly, as improvement occurs the dose can be reduced to once a week and still later, the patient should be maintained for life with once a month injections.

CHAPTER - V

INFECTIONS OF THE NERVOUS SYSTEM

Infections may reach the central nervous system via multiple routes. It may result from direct spread as in osteomyelitis of the skull or vertebrae, in mastoiditis, or sinusitis. Infection may come via the blood stream (haematogenous) or spread along the anastamotic veins that connect the facial and intracranial venous channels. Penetrating brain wounds can introduce infection and lastly infection may be introduced iatrogenically during a lumbar puncture using unsterile equipment, or during surgery. The iatrogenic causes of infections are fountunately very rare.

A variety of organisms can cause infection of the nervous system :

Bacteria The commonest bacterial infections are caused by meningococci, pneumococci, haemophilus influenzae and mycobacterium tuberculosis. Rarely staphylococci, streptococci, and pseudomonas infect the nervous tissue. Patients on immunosuppressive drugs and patients with carcinoma are more likely to fall prey to infections from rare organisms.

Viruses Common viruses infecting the central nervous system are Entero viruses like the Echo virus, Cocksackie virus, polio virus, Influenza virus A and B, meningoencephalitis complicating infectious fevers like measles, mumps, the variola-zoster group, the Epstein - Barr virus, and Herpes Simplex.

Spirochaetes Syphilis and Leptospiral meningitis

Fungi and yeast Cryptococci, Actinomycetes, aspergilosis, mucormycosis and Torula.

Parasites Malaria, cysticercosis, amoebiasis and Trypanosomiasis. Inflammation can also occur due to exposure to chemical agents, and after infiltration by malignant cells.

When an inflammatory process involves the pia and arachnoid membranes the resultant condition is called *leptomeningitis*, whereas in inflammation of the dura alone is called *pachymeningitis*. The term meningitis is generally used to mean leptomeningeal involvement.

MENINGITIS

The leptomeninges may be infected by any of the agents listed above. However, certain organisms are commoner in specific age groups, for instance *E. Coli* is the likely organisms in neonates, haemophilus in young children and pneumococci in adults.

Meningococcal meningitis It occurs in infants and very young children, and is said to be commoner in the early months of the year. Infection reaches the nervous system via the nasopharynx and is transmitted by human contacts. A septicaemia heralds the onset of the disease, there is high fever, malaise, and severe headache. Purpuric rash and the Waterhouse–Friderichsen syndrome, which results from haemorrhage in the adrenal glands following an intravascular coagulopathy may occur during this stage. Meningitis is heralded by intense headaches, restlessness, irritability, delirium and seizures. Herpetic eruptions may be seen around the mouth. Metastatic infection involving the eyes – uveitis, choroiditis, the bones – purulent arthritis, the heart – myocarditis, may complicate the course of the illness. The temperature is elevated, ranging from 39 to 40 degrees centigrade. Marked stiffness of the neck, and a positive Kerning's and Brudzinski's sign are found on examination. The reflexes may be exaggerated. Adhesions of the leptomeninges may obstruct the CSF pathway leading to hydrocephalus. The optic and auditory nerves may be severely damaged as a result of the infection. Chronic meningococcal septicaemia may also be complicated by a meningeal infection. Purpura, arthritis and low grade fever accompany the condition that results from a deficiency of the IGM immunoglobulin complex.

Meningococcal meningitis can be differentiated from other forms of meningitis by the isolation of meningococci from CSF. Pneumococcal meningitis is diagnosed demonstrating pneumococci in the CSF. Gram negative bacilli are seen in Haemophilus infection. The CSF shows similar findings in all types of purulent meningitis. The opening pressure is increased, the fluid is turbid, and has a polymorphonuclear pleocytosis. Protein content is markedly increased, whilst glucose is decreased and may even be zero. The chloride level depends upon serum chloride and is lowered if there has been repeated vomiting.

Meningococcal meningitis is treated by the use of benzyl or crystalline penicillin in a dose of 1 mega unit/kg/day intravenously in

divided doses. In adults the dose of penicillin is 24 to 30 mega units per day. Pneumococcal meningitis is also treated by benzyl penicillin in similar doses. Chloramphenicol in a dose of 100mg/kg/day in divided doses, is used in patients who are allergic to penicillin.

Haemophilus influenzae It is common in children below the age of 5 years, and has a subacute onset compared to meningococcal meningitis. The disease may begin with a nasal catarrh, bodyache, and fever with chills. Meningeal infection leads to drowsiness, irritability and headaches. Signs of meningeal irritation are present on examination. CSF findings are similar to those listed for purulent meningitis, and the Gram negative bacillus of *H. Influenzae* can be demonstrated on smear and culture. A common complication of *H. Influenzae* infection is a subdural effusion, which must be suspected when the child fails to improve after 72 hours of antibiotic therapy. Repeated tapplings may be necessary to evacuate the collection of purulent material. If a thick membrane has formed, surgical excision of the membrane becomes necessary.

Chloramphenicol in a dose of 75 mg per kg per day in divided doses is the treatment of choice. Ampicillin, 400 mg/kg/day can also be used.

Tuberculous meningitis This particular meningeal infection is rampant throughout India and may account for 5 to 8 % of all infections seen in this country. Tuberculosis can affect the nervous system in several ways, as meningitis, as a tuberculoma, as arachnoiditis, and as a infection of the vertebrae—Pott's disease—which may cause compression of the spinal cord and paraplegia.

Tubercular meningitis can occur at any age though it is commoner in children. The infecting organism is usually of the human type. Rarely the bovine or atypical form of microorganism invades the nervous system. Tuberculous meningitis usually occurs following tuberculosis elsewhere in the body. The meninges may be covered with disseminated miliary tubercles or focal plaque may proliferate leading to granulomas and adhesions.

A vague history of illness, and apathy is reported. Irritability, lack of appetite, and low grade fever are classical symptoms which are often missed or ascribed to other causes. In over 50% of cases the onset is acute, the patient presenting with fever, headaches, vomiting and

having had several seizures. In adults the onset may be subacute, with a history of confusion, loss of memory etc., or focal signs such as a mono or hemiparesis. Rarely a child is brought to the hospital in coma, with signs of acute hydrocephalus, and rapid deterioration. If the diagnosis is missed in the early stages, irregular remittent fever, drowsiness, irritability, and confusion occur. The CSF shows a lymphocytic pleocytosis, and a glucose level which though lower than normal, is not as low as in purulent meningitis. Very early cases of tuberculous meningitis may show a polymorphonuclear preponderance, and may be mistaken for purulent meningitis.

Tubercular meningitis may lead to focal neurologic deficits, and has a tendency to involve the basal regions of the brain, causing basal cranial nerve palsies. Spinal arachnoiditis may lead to radiculo-myelitis. If the brainstem is involved following arteritis in the basilar territory, drowsiness, pupillary abnormalities, ophthalmoplegias, and dysphagia may occur.

The course of the disease is radically altered by the prompt administration of antituberculous therapy. The CSF examination usually allows a diagnosis to be reached. Demonstration of tuberculous bacilli on smear, culture, or after guinea-pig inoculation confirms the clinical suspicion. Unfortunately these tests often cannot be performed in many hospitals in our country, so that cases of tuberculosis are treated on the basis of clinical suspicion and CSF findings alone. Patients with atypical histories or with focal neurological signs may need further radiologic investigations, including cerebral angiography to detect a tuberculoma. At times, arteritis at the base of the brain is demonstrated on angiography.

Treatment consists of the administration of three or four antituberculous drugs at the onset. Streptomycin, 40 mg/kg in children and 1 to 2 grams in adults. Isoniazid, 4 mg/kg in children and 300–400 mg/day in adults with paraaminosalicylic acid, 0.2 g./kg in children and 10 to 12 gms per day in adults or with pyrazinamide 20 to 35 mg/kg daily are the cheapest and most practical drugs in the treatment of tuberculosis in India. Thiacetazon, 3.4 mg/kg in children and 150 mg in adults may be added to the regimen if paraaminosalicylic acid produces side effects or the progress is not satisfactory. Recently, ethambutol and rifamycin have been added to the antitubercular armamentarium, but their prohibitive cost in India, limits their use to very severe and complicated cases. Steroids are

generally administered simultaneously as they help to reduce inflammation and the risk of adhesions, and also improve the general state of the patient especially in terms of a feeling of well being and an increased appetite. All 3 drugs should be continued for a period of three months and CSF examinations carried out every 2 to 3 weeks to monitor the progress of the disease. If a satisfactory response is not observed in the first 3 weeks, the diagnosis should be doubted, or the presence of complications suspected. A rise in CSF protein levels should lead to a suspicion of adhesive arachnoiditis and hydrocephalus. A ventriculo-peritoneal shunt may have to be inserted the patient may otherwise lapse into a coma. The use of intrathecal Hyalase (1500 units) every 15 days has been suggested recently to treat adhesions especially in the spinal form of the disease.

SPINAL ARACHNOIDITIS It can be one of the common complications of, or the only manifestation of, tubercular meningitis. It may present subacutely or in a chronic form. The subacute form begins with radicular pain that resemble the pain of sciatica. The pain may involve single or multiple roots. The root involvement gradually ascends, and the pain radiates down the arms. Pain varies from a dull aching to a lancinating character. As the spinal cord may also be involved a spastic paraplegia or even quadriplegia may be seen. Due to root and cord involvement a mixed type of paralysis is seen the upper limbs. There is wasting and weakness of the small muscles of the hands. Fasciculations may be observed in the deltoids, biceps, and the flexors of the forearms. There is inversion of the supinator reflex, the reflex itself being absent, with exaggeration of finger flexion. Sphincter involvement occurs early in the disease Paraesthesias and dysesthesias are common, starting at the sacral level and ascending to the trunk. On examination there is proprioceptive loss in the lower limbs.

The CSF is xanthochromic with a lymphocytic pleocytosis, and a marked increase in protein. The glucose is low in the tubercular variety of arachnoiditis. Syphilis can cause a similar symptom complex, and is diagnosed by the positive VDRL reaction and Wasserman test in the CSF. On myelography 'candle guttering' is seen, with multiple levels of blocks and fragmentation of the dye column so characteristic of this disease.

Treatment is largely medical, with antituberculous drugs and steroids being administered in the tubercular variety, and steroids and

penicillin in the syphilitic variety. 1500 units of Hyalase is injected intrathecally fortnightly, and if improvement is noted, treatment should be continued till no further improvement occurs.

NEUROSYPHILIS

The incidence of neurosyphilis has declined dramatically since the advent of penicillin, a medical student often does not get the opportunity to study a patient with this disease whilst on the clinical services.

The primary chancre is generally the first manifestation of acquired syphilis, the treponema being already disseminated in various tissues of the body, with central nervous system involvement occurring in the second stage of the disease. The involvement is asymptomatic, but meningovascular syphilis may present at this stage. Meningovascular syphilis can be divided into two forms: the cranial and the spinal form. The cranial form includes syphilitic meningitis, a 'stroke' like syndrome giving rise to hemiplegia, gumma, optic atrophy or nerve deafness. The spinal form consists of meningomyelitis, cervical pachymeningitis, syphilitic amyotrophy and radiculitis. Symptoms may develop a few months to a few years after the primary infection and depend on the site of involvement of the meninges or blood vessels.

Acute syphilitic meningitis behaves like any other acute meningeal infection presenting with fever, headaches, vomiting, signs of meningeal irritation, and polymorphonuclear pleocytosis in the CSF. The protein is increased and the VDRL is strongly positive. The chronic variety presents with involvement of a single, or multiple cranial nerves, with the third, fourth, sixth and eighth being commonly affected. Symptoms of a meningeal infection are present, including mental changes. Adhesions at the base of the brain lead to hydrocephalus and signs of increased intracranial pressure. If the meningitis is restricted to the convexity of the brain, headaches and focal seizures occur. The CSF shows a moderate lymphocytic pleocytosis, with a normal glucose, a minimal protein elevation, and a positive VDRL.

The typical vascular lesion of syphilis is an endarteritis, with the middle cerebral artery being commonly affected, producing a contralateral hemiplegia. Signs of increased intracranial pressure and presence of focal neurologic deficit should lead to a suspicion of a syphilitic 'gumma'. The gumma is rare, and is associated with a positive VDRL in the CSF. Adhesive arachnoiditis, gumma formation, and endarteritis of

the vasa nervorum can lead to lesions of the optic nerve and chiasm. Symptoms consist of progressive loss of vision in one eye with preservation of central vision, and marked pallor of the disc on ophthalmoscopic examination. Chiasmal lesions produce field defects with a bitemporal loss of vision.

The commonest form of meningovascular syphilis is the spinal form with meningomyelitis. The course is subacute, backache and root pains in the dorsal region being the first symptoms. A spastic paraplegia develops with sphincter involvement and a thoracic sensory level. Sometimes an anterior spinal artery syndrome occurs, with preservation of joint-position sense. If the posterior spinal arteries are also involved, a transverse myelitis may be seen. Erb's syphilitic paraplegia occurs 5 to 10 years after the primary infection. The symptoms are those of a gradually progressive spastic paraplegia, with bladder involvement and impotency. Sensory changes are mild or absent. A rare form of syphilis is the amyotrophic form which resembles amyotrophic lateral sclerosis. The CSF VDRL is positive and treatment with penicillin arrests the course of the illness.

In all forms of meningo vascular syphilis treatment consists of administering large dose of penicillin. Recently intravenous penicillin in doses similar to those used for purulent meningitis have been recommended. This form of treatment is continued for 10 days and has the best results. Treatment should be repeated in 6 to 8 weeks if there is no significant improvement in the clinical status or CSF abnormalities remain unchanged. Remember it is the presence of cells in the CSF that indicates active disease, as the protein remains high for months to years, and the VDRL may never revert to the negative state.

General Paralysis of the Insane (GPI) This is parenchymal form of syphilis and occurs 5 to 25 years after the primary infection. It is more frequent in males than in females. The onset is insidious, the patient presenting with illdefined symptoms such as headaches, restlessness, insomnia and loss of drive. Changes in personality and alteration in mood and behaviour are reported. As the disease progresses deterioration in memory, loss of judgment and lack of social inhibition may be seen. A flat affect and lack of appropriate emotional responses, have also been noted. Euphoria and grandiose delusions with blunting of affect are observed in the "grandiose form of GPI". Seizures and attacks of transient neurologic deficits occur in the later stages, the patient being totally demented and requiring institutional care at this stage.

Physical signs include a facies that shows no expression. A fine tremor affects the tongue, lips and fingers. Optic atrophy is seen in about 10% of patients. The pupils show the classical Argyll-Robertson abnormality: small, irregular pupils that do not react to light, but react promptly to accommodation. The iris may appear to be muddy in colour. The speech is dysarthric, and the limbs show signs of bilateral pyramidal tract involvement with hyperreflexia and extensor plantar responses. The CSF shows a mild to moderate increase in cells (Lymphocytes), an elevated protein level and a "paretic" gold curve (a first zone rise). The VDRL is positive. The Colloidal gold curve is a rough index of increased gammaglobulin in the CSF. Ten tubes of CSF are tested in varying dilutions against constant amounts of colloidal gold solution. Changes in the form of precipitate formation are recorded on a scale of 1 to 5, with 0 representing no change and 5 denoting maximal precipitate. A 'paretic' curve or a first zone rise is therefore associated with precipitate formation in the first four or five tubes and is numerically expressed as 5555321000. A 'tabetic' curve seen in *tabes dorsalis* gives a mid zone rise: 123321000 and a 'meningitic' curve shows a further shift of the right: 0012344310. The colloidal gold curve has lost its preeminence today, as direct measurement of CSF immunoglobulins is technically feasible. However, the test is still useful in the diagnosis of neurosyphilis and Subacute sclerosing panencephalitis. (SSPE).

GPI should be treated in a manner similar to syphilitic meningitis. If no improvement occurs a quaint form of therapy that induces fever either by the use of the parasite of benign tertian malaria or by injection of typhoid vaccine etc. has been found to have beneficial effects.

Tabes dorsalis It is another parenchymal form of syphilis and has a long latent period, occurring 10 to 15 years after the primary infection. Like GPI, it is commoner in males and occurs around the fifth decade of life. The lesion is in the dorsal nerve roots. There is degeneration of the posterior columns with meningovascular involvement of the dorsal root ganglia and dorsal nerve roots. Fibres subserving pain are first to be involved, leading to the characteristic lightning pains of tabes. They generally occur in the distribution of the sciatic nerves. Root involvement at the thoracic level results in girdle pains. The pain varies in intensity, lasts for a short time, and shifts from site to site. The patient describes the pain as "though needles were piercing the limb in a perpendicular direction". Pain may affect the viscera causing the typical 'tabetic crises'. The viscera involved include the stomach, the larynx, the rectum and the bladder.

As the disease progresses, there is loss of deep sensation and squeezing the patient's calf or testes causes no pain. The patient complains of difficulty with gait that is worse at night. This is due to proprioceptive loss, the Romberg's sign being positive at this stage. Loss of the reflex arc occurs, and typically the ankle jerk is the first reflex to be lost. Marked hypotonia leads to extreme mobility of the joints. The sensory loss and the hypotonia together contribute to the progressive destruction of the joint: the classical deformed 'Charcot's joint'. Total destruction of the nerve roots leads to the 'high stamping gait' where the patient lifts his knee high off the ground, and being unable to judge the distance between the foot and the ground, bring the foot down hard, stamping it into the ground. The patient may state that he feels as if he is walking on cotton wool.

On examination, optic atrophy may be present, Argyll-Robertson pupils are seen and there may be diminution of sensation in the 'butterfly area' of the face, that is the cheek area. Anaesthesia or hypoaesthesia may be demonstrated over the inner border of the forearm, the "saddle area" of the buttock, and the feet. There is marked hypotonia, with loss of the ankle jerks. The plantar response is flexor, though extensor responses may be seen in cases of tabo-paresis. A profound proprioceptive loss, and a positive Romberg's sign are seen. The high stamping gait may be demonstrable and Charcot's joints may be observed. Perforating ulcers on the soles of the feet may be seen. Bladder incontinence with dribbling of urine, a large bladder and a large volume of residual urine are found. There is total loss of bladder sensation. Impotence is common.

The CSF shows a mild lymphocytic pleocytosis of a 100 to 150 cells, with a second zone rise in the colloidal gold curve the "tabetic curve". The signs and symptoms of tabes are so typical that diagnosis presents little difficulty. The presence of optic atrophy and the pupillary abnormalities serve to differentiate tabes dorsalis from other diseases of the spinal cord and the nerve roots. The treatment is similar to other forms of neurosyphilis.

The student would do well to remember the aphorism that syphilis is the "great imitator" and that the disease has protean manifestations. Syphilis should be a part of the differential diagnosis of any neurologic disorder of acute or chronic nature, especially in isolated cranial nerve palsies or spinal cord disease. The student of medicine in India must never forget the social milieu in which she or he has to practice.

The term 'encephalitis' generally means a non-suppurative inflammation of the brain. Many viruses affect the nervous system, the common ones being rabies (Rhabdo virus), the virus of Japanese B encephalitis (Arbo virus), the polio virus, the coxsackie and the Echo virus, and the viruses of measles (Myxo), smallpox (variola) and the viruses causing herpes simplex and herpes zoster infection. Of these rabies, Japanese B and enteroviruses infect the nervous system directly, whilst CNS involvement in mumps, measles, variola, herpes and varicella is secondary to a generalised infection that may rarely involve the nervous system.

JAPANESE B ENCEPHALITIS : In recent years multiple epidemics of this infection have occurred in India, especially in the northern, eastern, and central regions of the country. Another Arbo virus that is common to south India is the Kyasanur Forest disease virus. Birds, especially pigeons are the primary host of Japanese B encephalitis, whilst monkeys are the hosts for Kyasanur forest virus. Mosquitoes are the vectors for Japanese B virus, whilst fleas, ticks, and mosquitoes are the vectors for Kyasanur forest virus. In the initial stage of viraemia the patient complains of fever, bodyache, lethargy, and generalised weakness. Headaches, vomiting, restlessness, irritability, delirium, and drowsiness herald the onset of the encephalitis. Frequent seizures occur, and coma ensues. Behavioural disturbances and thought disorders may occur in the early stages of the illness, leading to an erroneous diagnosis of a psychiatric disease. Focal neurological signs in the form of a monoplegia or a hemiplegia may be present. Signs of meningeal involvement are minimal or absent.

CSF examination shows a mild lymphocytic pleocytosis, with minimal elevation of protein. The glucose is normal. The EEG may show generalised slowing or focal or generalised epileptiform discharges. The diagnosis is relatively simple when an epidemic rages. In sporadic cases, the diagnosis is made by the history, the lack of meningeal involvement, and the typical CSF findings. Toxic encephalopathies can be differentiated by the history of exposure to toxin, and metabolic encephalopathies are associated with features of the primary disease like uraemia, and liver failure. Brain abscess may be difficult to rule out in some instances, and invasive procedures including cerebral angiography may be needed before a firm diagnosis can be made.

Treatment is largely, symptomatic, with steroids providing dubious benefit. Hyperthermia, restlessness and irritability, and seizures should

be treated with appropriate measures. Good nursing care is imperative to prevent bedsores. The morbidity of encephalitis is high, with chorea, extrapyramidal problems such as a Parkinsonian syndrome, sleep disturbances, epilepsy, intellectual deficits and hemiplegia being prominent sequelae.

Post infectious encephalitis shows similar clinical features, and occurs either during the course of the infectious fever or rarely, immediately after the infection. Encephalitis, encephalomyelitis, and peripheral neuropathies may occur with measles, mumps, variola and varicella infections.

Herpes simplex encephalitis is the commonest non-epidemic form of encephalitis in the West, but has been reported only rarely in India. The virus has a predilection for the temporal lobe, behavioural disturbances and seizures are common, as are focal neurologic signs. There is massive swelling of the temporal lobes, and consequent rise in intracranial pressure. Isolation of virus by immunofluorescent techniques is diagnostic, and prompt administration of Adenine Arabinoside, an antiviral agent, reduces mortality from 70% to around 30%.

Poliomyelitis Despite the availability of polio vaccine, the disease continues to scar children in the Indian subcontinent another public health failure that serves to highlight the medical aspects of faulty social priorities and consequent neglect of public health. The authors hope that students will not merely attribute the occurrence of diseases such as poliomyelitis to poverty and ignorance but analyse reasons for the persistence of these diseases, decades after the subcontinent has achieved remarkable progress in other industrial and medical field. Pathologically the lesions involve mainly the anterior horn cells of the spinal cord and the cells of bulbar cranial nerve nuclei.

The illness consists of four phases : 1) a minor illness that may consist of a mild diarrhoea. 2) the prodromal illness consisting of fever, malaise, coryza, and gastrointestinal disturbances for 2 to 3 days. The symptoms then abate for a few days before the **third** stage, or the pre-paralytic stage, begins. The temperature rises, the patient has a severe headache, and stiffness of the neck. There may be severe pain in the limbs, and even minimal movement of the limbs causes excruciating pain. The patient therefore resists any attempt to move the limbs. The paralytic stage, the **fourth** stage, sets in, the pain abates, the fever drops,

and constitutional signs regress. Motor paralysis, that is asymmetric, and may be localised or wide spread then becomes marked. Affected muscles are flaccid, weak, and atrophic. Tendon reflexes are lost in the affected muscles. There is no sensory loss, and fasciculations are rare. In some patients the paralysis spreads to involve the bulbar musculature, causing respiratory distress. The patient may then need assisted ventilation, and requires intensive medical care. A tracheostomy may be necessary, and needless to say, patients with bulbar paralysis have a high mortality and morbidity.

The formation of contractures should be prevented in the convalescent stage, with intense rehabilitative measures offering the best hope for the patient. The CSF shows a polymorphonuclear pleocytosis in the first week, later changing to lymphocytic preponderance, with moderate elevation of protein and normal glucose levels. The virus can be isolated from the stool, serum and CSF. The viral antibody titres rise dramatically and acute and convalescent titres should be studied. The acute titre studied on admission, and the convalescent titre studied after 2 to 3 weeks.

The diagnosis should not be confused with rheumatic arthritis, osteomyelitis, scurvy, and acute infectitious polyneuritis. Careful examination of the bones and joints, and presence of swollen bleeding gums, should be noted. Acute infectious polyneuritis is differentiated by the symmetric distribution of weakness, the presence of unilateral or bilateral facial involvement, the presence of some sensory deficit, and the absence of cells in the CSF with a massive elevation of the protein level.

In 1971 and 1981 epidemics of acute haemorrhagic conjunctivitis have occurred in India. Extensive polio like paralysis have been reported as sequelae, the complication being rather frequent in the epidemic of 1981. Males were more commonly affected, and there was no age preference. In some cases cranial nerves, especially the seventh nerve were involved.

Management Strict bed rest is imperative in the prodromal preparalytic and paralytic stages. Intramuscular injections should be strictly avoided and light splints used to immobilise a limb. The judicious use of sedatives and analgesics does much to relieve the patient's distress. The pain which is due to muscle spasms may be caused by local measures like

hot fomentations. After the pain subsides, passive movement and later active physiotherapy should be started. Corrective surgery, tendon transfers, use of braces etc. must be considered in the rehabilitation of the patient.

The disease is now preventable and oral polio vaccine given at 6 months of age and repeated after 6 weeks and 6 months effectively prevents this disease. A booster dose may be given when the child enters school and at 15 or 20 years of age.

With evolution of concept of slow virus infection from the study of the disease called 'Kuru' in New guinea, two more slow virus diseases in humans have come to light. Subacute sclerosing panencephalitis is common in India, while, Jakob-Creutzfeldt disease is extremely rare.

Though the incidence of Subacute sclerosing panencephalitis in western countries, in on the decline, it is still frequently seen in India. The disease occurs around 6-14 years of age. There is history of measles infection in early childhood, the infection then persists. It begins with gradual intellectual deterioration. Few months later patient develops myoclonic jerks and convulsive episodes. From then on he progressively goes downhill, with marked mental deterioration, aphasia, apraxia, and quadraplegia, leading to a vegetative state and death, within few months or years of onset of the disease. High measles antibody titres in the serum and CSF have been reported. The EEG shows characteristic recurring periodic discharges. As yet there is no known treatment.

Jakob-Creutzfeldt disease is a disease of the central nervous system with a subacute progressive course. The infection is transmissible to animals. It presents with progressive dementia, and myoclonus, which are the prominent clinical features. On examination, evidence of pyramidal and extra-pyramidal systems involvement is obtained. Signs of lower motor neurone involvement appears in the later stage of the disease. Once again the EEG shows characteristic periodic generalized sharp waves recurring every $\frac{1}{2}$ to 2 seconds. Patient usually dies within six months of onset of the disease.

CYSTICERCOSIS It is seen throughout the country but is maximum in northern and southern states of India. The causative agent is *cysticercus cellulosae*—a larval form of tape worm—*taenia solium*. The infection develops by eating inadequately cooked, infected pork or contaminated

food. It may present in a variety of ways. Patient may present with epilepsy, or with evidence of encephalitis, meningo-encephalitis or signs of increased intracranial tension. Multiple small nodules may be detected elsewhere on the body and biopsy of one of them will reveal presence of cysticercus. Cysts calcify after 4-5 years and radiological examination of the skull may show calcification in some of the cysts. The treatment is symptomatic.

CEREBRAL MALARIA Malaria, once extinct from India, is seen once again. Infection with *Falciparum* strain, can cause cerebral malaria. Diagnosis of cerebral malaria is not easy, and is made on presumptive evidence. If a patient with malarial fever develops coma or signs of confusion, restlessness, convulsions or focal neurological signs and has positive blood smears for *plasmodium falciparum* infection, a diagnosis of cerebral malaria be made and the patient so treated. Prompt response to therapy would further substantiate the diagnosis. However, exclude other causes of coma or encephalopathy at the same time.

The treatment consists of intramuscular injection of 200 mgm of Chloroquine base (250 mgm as hydrochloride), half the dose is given in each buttock. This may be repeated every 6 hours and in first 24 hours the dose should not exceed 800 mgm of chloroquine base. Intravenous infusion of glucose saline should be started. Dexamethasone is recommended to reduce the brain oedema. If the *falciparum* strain is resistant to chloroquine, quinine should be given in the infusion and a watch kept on the pulse and blood pressure.

CHAPTER – VI

EPILEPSIES

The word epilepsy is derived from the Greek words 'epi' meaning upon and 'lepos' meaning to seize. In ancient times an attack of seizures was believed to be due to the patient's soul being seized by a demon. Hippocrates discounted these myths and was the first to observe that seizures originated from the brain. Hughlings Jackson laid the foundation for the modern understanding of epilepsy. He published a series of brilliant papers in the nineteenth century, and delineated the different types of seizures. The introduction of electroencephalography (EEG) helped to understand the basic electrophysiologic mechanisms of epilepsy and recently the use of positron emission tomography (PET) promises to unravel further the sequence of events in a given seizure.

It is important that the student realise at the onset the difference between the terms "seizure" and "epilepsy". A seizure is the result of a 'sudden, excessive, rapid discharge' of neurones. Epilepsy, on the other hand, is the occurrence of two or more seizures, that do not occur in relation to drug withdrawal or to an acute illness. Febrile seizures are seizures that occur between the ages of one month and 6 years, the first seizure ever experienced was accompanied by a fever, and the seizure is not a symptom of a recognised acute neurologic illness. The prevalence of epilepsy is said to be 6 per thousand, though the figure is higher in poorer populations. The role of inheritance in epilepsy remains controversial, though certain facts are accepted by all workers in the field. When the epilepsy is of the Absence type, there is an 80% chance of the siblings demonstrating the typical spike and wave pattern on the EEG. As yet there are no suitable controlled studies that identify the risk of developing seizures or epilepsy and the final word on genetic counselling is awaited. The causes of seizures are tabulated below.

TABLE 6.1
CAUSES OF CONVULSIONS

Infections	Meningitis—specially tuberculous meningitis in our country. Encephalitis, Brain abscess, Tuberculomas and parasitic infestation like Cysticercosis.
Trauma	Concussion, Contusion, Penetrating head injuries, Subdural Haematoma.
Toxins	Heavy metals like arsenic, lead, organo-phosphorus and organochlorine compounds used as insecticides, Carbon monoxide poisoning, Alcohol, Drugs : Imipramine, Isoniazide, Cardiazol, Nikethemide.
Metabolic	Hypoglycaemia, Uraemia, Hypoxia, Alkalosis, Pyridoxine deficiencies, Hypocalcaemia, Porphyrrias, Eclampsia, Phenylketonuria.
Tumours	Primary and Secondary
Vascular	Cerebral infarction, Subarachnoid haemorrhage Hypertensive encephalopathy, Stokes-Adams syndrome, Systemic lupus erythematosus, Arterio-venous malformations.
Congenital anomalies	Cerebral diplegia, Hydrocephalus,
Degenerations	Freidreich's ataxia, Ramsay Hunt syndrome, Lipid storage disorders, Multiple sclerosis and Schilder's, Picks & Alzheimer's disease.
Miscellaneous	Fever, Alternate light & shade effect, Television etc.

Seizures are classified according to the International Classification of Seizures. They are divided into two broad categories: Generalised seizures and Partial seizures.

Generalised seizures are those seizures in which the onset is bilateral and results in impairment of consciousness. The main types of generalised seizures are:

- 1) Absence seizures
- 2) Tonic-clonic seizures
- 3) Myoclonic seizures
- 4) Atonic seizures

Partial seizures are those seizures in which the seizure begins locally either clinically or on the EEG. Partial seizures are further classified into Simple partial seizures where consciousness is **not** impaired during the seizure, and Complex partial seizures where there **is** impairment of consciousness during the seizure.

We shall now describe the common types of seizures, and define certain terms that are frequently used in epilepsy.

ABSENCE SEIZURES (PETIT MAL) These seizures are common in childhood, with the age of onset being 5-7 years. They comprise between 3-10% of all seizures but are less common in India than in Western countries. The seizures begin and end suddenly. There is no aura, no massive spasms, no formed speech and no post ictal phase. The average duration of an absence is ten seconds, with spells longer than 45 seconds being very unusual. The simplex absence with just impaired responsiveness occurs in less than 10% of absences. The most common spell involves mild clonic activity and simple automatisms like lip licking and lip smacking. *Automatisms* are involuntary motor acts that occur during a seizure and are followed by amnesia for the event.

Absences are relatively easy to treat, with ethosuximide (Zarontin) and Valproic acid being drugs that are very effective in controlling the spells. The frequency of absences varies between 25 to over a 100 seizures a day. Patients may have generalised tonic-clonic seizures in addition. The EEG shows the typical 3 per second spike and wave discharge (Fig 6.1). The electrical abnormality also begins and ends abruptly like the clinical attack. Hyperventilation almost always precipitates an attack of absences. The prognosis for remission from these seizures depends upon the normalcy of mental development and neurologic examination, the typicality of the EEG findings and the absence of generalised tonic-clonic seizures, and a negative family history of seizures. If all 4 factors are present, there is an 80% chance of remission by the time the patient has reached puberty.

ictal phase which may consist of drowsiness, confusion, difficulty with speech, or muscle soreness and fatigue. Headaches are also a common post ictal phenomena. Paralysis of a limb following a generalised tonic-clonic seizure, with recovery from the paralysis within 24 hours is known as 'Todd's paralysis'. The patient may fracture his thoracic vertebra during the tonic phase of the seizure. Some patients injure themselves very frequently after their spells, and many of them unfortunately belong to the small group of patients whose seizures are refractory to medical therapy. A few patients have a prodrome of changes in mood, or muscle twitches a day or so before the seizure. The frequency with which patients may have generalised tonic-clonic seizures varies from 2-3 per year to several in a single day. Seizure frequency can be reduced by appropriate medications in the majority of patients. Certain situations like infection, periods of severe stress, and metabolic imbalance can cause an increase in seizure frequency. Noncompliance with prescribed drug regimen is the most common cause of seizures in patients who are otherwise well controlled. Only 25% of individuals with a chronic non-painful illness follow drug regimens accurately, so that non-compliance is a fairly common problem in any large seizure clinic.

The EEG shows a rapid train of spikes during the tonic phase of the seizure. It changes to polyspikes and slow waves during the clonic phase, and shows diffuse slowing in the post ictal period. The post ictal changes may persist for one to two days after a seizure. An inter-ictal EEG will show abnormality in 40% of patients. These abnormalities include generalised spikes (Fig 6.2), focal spikes, and spike and wave or localised sharp slow wave complexes. A spike is a wave of very high frequency, and a sharp wave has a duration of less than 70 milliseconds. The positivity of epileptiform abnormalities increase with repeated recordings, the induction of sleep, a short duration between the EEG recording and the last seizure, and the use of activation procedures like sleep deprivation, hyperventilation, and photic stimulation. Special electrode placements such as the nasopharyngeal electrodes increase the possibility of demonstrating epileptiform discharge especially in patients with complex partial seizures.

Generalised seizures have no known cause in 60% of cases. Head injury, perinatal anoxia, strokes and brain tumours are some of the known causes. Arterio-venous malformation, granulomas, and toxic conditions like lead encephalopathy are other conditions that cause generalised seizures.

The diagnosis of epilepsy is a *clinical* diagnosis and should be made on the basis of a careful history. Question the patient closely about the nature of the spells. When did they first begin? Did the patient have problems with seizures as a little baby? Did he have seizures with fever as child? Did he ever lose consciousness as a result of a head injury? Did he ever have meningitis or encephalitis?

What did his first seizure consist of? Does he have a warning immediately preceding the seizure (aura)? Does he remember what had happened during the spell? Has anybody observed the spell and did they tell him what happened to him during the spell? Do his limbs jerk, does he stiffen, does he chew his tongue? Does he pass urine? Does he injure himself? How long does the spell last? Does he come back to normal at once, or does he feel sleepy and confused after the seizure? How often does he have seizures? Do they occur at night? Do they occur when he is alone? Does drinking alcohol increase his seizures?

A new case of epilepsy must be investigated with at least an EEG, when indicated a C.T. brain scan, to search for a focal lesion that may be responsible for the seizure. Psuedoseizures (hysterical) may mimic generalised seizures, and the problem is further complicated as the two may coexist in the same individual. Caution is advised before diagnosing pseudoseizures, as the novice may be deceived by a rare seizure type like the supplementary motor area seizure. When in doubt admit the patient and observe closely the actual seizure. The table below will help to differentiate most psuedoseizures from true seizures.

TABLE 6.2

DIFFERENTIATING FEATURES OF HYSTERICAL AND TRUE SEIZURES

Hysterical Fits	Epileptic seizure
Induced by emotional upsets.	Needs severe emotional stress for precipitation
Irregular periodicity as it is precipitated by minor upsets.	Fairly regular periodicity.
Never in sleep	Can occur during sleep

No incontinence, injury, tongue biting	One or all can occur in one of the attacks.
Always in presence of people, bizarre spectacular movements, corneal reflex preserved	Can occur in isolation, tonic-clonic movements absent.
Eyes generally closed and resisted if opened, fists clenched tight,	Eyes rolled up and fists not clenched.
Spectacular recovery-hyperventilations, sighing, yawning etc.	Gradual recovery.

Treatment Most patients with epilepsy should continue normal activities such as schooling and working, though care must be taken that the patient does not find himself in a situation where he would injure himself badly should he suffer a seizure. These instances include swimming alone, climbing heights, and being close to heavy machinery. Epileptics surprisingly have a lower accident rate than non-epileptics because they are more cautious in the workplace than their colleagues. Nothing should be done that makes the patient develop an inferiority complex or lowers his self esteem. In India legislation regarding the rights of epileptics is behind the industrialised countries, and this situation is badly in need of remedy. For instance epileptics are not allowed to drive in our country, whilst in the West an epileptic may drive if the seizures are well controlled with medications.

The medical therapy for generalised tonic-clonic seizures consists of choosing between any of the following drugs: phenobarbital, diphenylhydantoin (phenytoin), carbamazepine, valproic acid, and primidone. Valproic acid has just been marketed in India. Before we discuss the details of individual anticonvulsant drugs we must remember certain principles that govern the use of anticonvulsant drugs. These principles are:

- Use drugs that you are thoroughly familiar with and use well known drugs before trying out less well-known ones.
- Use one drug and raise the dose close to the toxic range before deciding that the drug is ineffective in seizure control.
- Drugs that have short half-lives have to be given in divided doses whilst drugs with long half lives may be given in a single dose.
- The time taken by a drug to reach steady state is generally five times the half life of the drug.

- e) Drugs with long half lives need a loading dose to reach steady state quickly.
- f) No drug should be discontinued suddenly as this may precipitate status epilepticus.
- g) Drugs used must be specific for seizure type.

Phenobarbitone is effective in all types of partial seizures and in generalised tonic-clonic seizures. It is both safe and cheap, its cost making it the most commonly used anticonvulsant in India, where the cost of medicines should always be a factor that governs physician's decisions. It is given in two divided doses in children, but may be given in a single daily dose in adults. A dose of 5 mg per kg is prescribed in children, with adults needing about 90-120 mg per day. The half life of phenobarbital varies between 3 to 5 days. Adverse side effects include drowsiness, skin rash, ataxia, and paradoxical hyperkinetic behaviour in children.

Diphenylhydantoin (DPH) or phenytoin (Dilantin, Eptoin): Is a first line drug for all types of partial seizures and for generalised tonic-clonic seizures. It is generally given in a dose of 300 mgms per day. A few patients may need a higher dose because of defective absorption or faster metabolism. The half life of DPH varies between 21-36 hours. Patients on chronic DPH therapy should be checked for ataxia, nystagmus and gum hypertrophy. Ataxia is a sign of toxicity and indicates that the dose of DPH needs to be lowered. Drowsiness and lethargy occur at higher doses.

Carbamazepine is also a first line drug in all types of partial seizures and generalised tonic-clonic seizures, but its use is restricted by its expense. It is ideal for children as it has neither the behavioural problems that may arise with phenobarbital, nor the cumulative side effects of DPH such as coarsening of the facial features, hirsutism, and aggravation of acne. Carbamazepine has a short half life of 6-8 hours with chronic use. It should be begun at a low dose and increased to a dose of 1400 - 1800 mgms per day. Its side effects are dose related, and may require many divided doses with a third of the dose being given at night. The side effects are ataxia, dizziness, blurring of vision, and diplopia. Leucopenia and even aplastic anaemia have been reported as rare side effects, and when evident require immediate discontinuation of the drug.

Valproic acid is a first line drug in absences, and in myoclonic seizures, and is useful in generalised tonic clonic seizures and atonic

seizures. It is given in divided doses, (half life of 6-8 hours) and may cause gastric irritation initially. Major side effects include those resulting from drug interaction (drowsiness when used conjointly with phenobarbital), ataxia, and weight gain. Hepatic dysfunction severe enough to cause death has been reported with the use of valproic acid and frequent liver function tests should be done whenever valproic acid is begun on a patient.

Other convulsants used include primidone (mysoline) and sulthiame (Ospolot). Ospolot is not available in India, and is sometimes useful in controlling generalised seizures. Mysoline causes drowsiness and ataxia, and is converted into phenobarbital in the body. It is given in doses of 250 mgms 3-4 times a day. Therapy should be initiated with very low doses like 125 mgms three times a day, otherwise the patient may develop signs of toxicity.

It is advisable to use a single drug and to increase the dose close to the toxic level. Only when there is inadequate seizure control with an appropriate drug specific to seizure type, should another drug be added. Cases that prove resistant to adequate doses of two or more anticonvulsant drugs may need intensive study, and should be investigated for possible surgery. Unfortunately surgical therapy is helpful in only a very small number of cases with complex partial seizures.

Myoclonic seizures Myoclonus can occur as an epileptic phenomenon or as a sign in a variety of degenerative, toxic and infectious conditions. As an epileptic phenomenon it occurs in young people aged 5-15 years. It consists of myoclonic jerks that occur nocturnally and may proceed to a generalised tonic clonic seizure. The EEG may show generalised spikes or poly spikes and waves (Fig 6.3). This seizure type is also known as the "benign myoclonic epilepsy of adolescence", and has a good prognosis. Treatment consists of DPH or valproic acid.

Atonic seizures These seizures are difficult to control and tend to occur in individuals with diffuse brain damage. The seizures consist of brief attacks of loss of tone, with the patient falling and injuring himself. The EEG show one to one and a half per second spike and wave discharge with slowing of the background. Valproic acid may be useful and in some cases ACTH therapy is effective.

Infantile spasms form a part of the West's syndrome. It consists of a triad of seizures, mental retardation and hypsarrhythmia on the EEG. Infantile spasms consist of brief spasms that last less than a second and

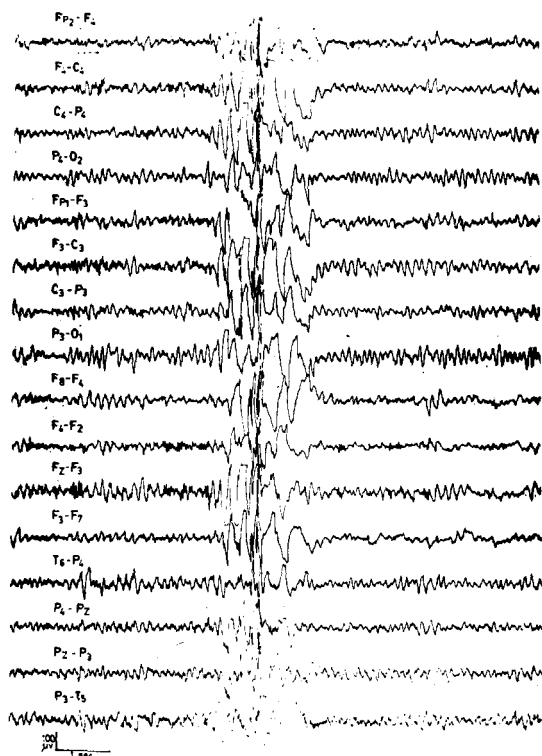


Fig. 6.3 : Generalized polyspike and wave discharge of myoclonic epilepsy.

most commonly are of the mixed type. The upper half of the body extends and the lower half flexes as if in obeisance to a medieval deity (Salaam seizure). The seizures have their age of onset prior to the end of the first year of life, and progress to become the minor motor seizures of childhood. The causes of infantile spasms include birth anoxia, phenylketonuria, tuberous sclerosis, and rarely following encephalopathy from pertussis vaccine. The prognosis is poor with over 90% of the patients being severely retarded. The EEG abnormality of hypsarrhythmia consists of a chaotic pattern, with polyspikes and mountainous slow waves. ACTH and valproic acid may be effective in this seizure type.

ACTH is given in doses of 20 to 40 units daily. Prednisone may be used if ACTH is not available or is too expensive, in doses of 1 mgm per Kg.

PARTIAL SEIZURES As explained previously, this includes two categories: Simple partial and complex partial, the difference being that consciousness is impaired in the latter but **not** in the former. We shall now discuss the main types of partial seizures.

1) With motor signs Any part of the body may be involved in this seizure, depending upon the site of discharge in the motor cortex. The Jacksonian 'march' is a partial seizure in which the seizure progresses in an orderly manner, with motor activity occurring in the thumb, the angle of the mouth, and the great toe. An adverse seizure consists of the head and eyes moving to the side opposite to the discharge, and sometimes progressing to a full blown generalised tonic clonic seizure. Occasionally aphasic seizures may be seen, the patient having difficulty with speech because of epileptiform discharge involving the speech areas. Focal seizure activity that continues for hours to months is known as 'epilepsia partialis continua' or Kojevnikov's epilepsy'.

2) With somato-sensory or special sensory symptoms Somato-sensory seizures arise from discharge in the sensory cortex. They consist of sensations of tingling described as "pins and needles feeling", or of a feeling of numbness. Rarely a disorder of proprioception or spatial perception occurs. Sensory seizures may also 'march' in the manner of motor seizures.

Special sensory seizures include visual seizures where hallucinations vary from flashes of light to vivid scenes being recalled, depending on the site of the discharge. Auditory hallucinations are rare in epilepsy, but common in psychotic conditions. Epileptic auditory hallucinations parallel the visual hallucinations in their elaborateness. They can vary for a single sound being repeated, to a whole song. Epileptic auditory hallucinations differ from those seen in the psychosis, because there is no thought disorder, and the patient recalls the hallucination with a sense of detachment.

Olfactory hallucinations are generally in the form of unpleasant undescrivable odours, the discharge originating in the uncus of the temporal lobe. Gustatory hallucinations parallel the sensations of taste, though commonly a 'metallic' or bitter sensation is described. Vertiginous symptoms include a sensation of floating in space, but these seizures are RARE and the reader is cautioned against calling any dizzy spell a vertiginous seizure.

3) With psychic symptoms These usually occur with complex partial seizures. They include dysamnesic symptoms such as *deja-vu*, a feeling of familiarity with an unfamiliar situation, and *jamais-vu* where

the patient feels strange in very familiar surroundings. Cognitive disturbances including dreamy states, distortions of time sense and feelings of depersonalization (autoscopy) may occur.

Seizures with affective symptomatology include those with either fear, pleasure, anger, and depression. These occur in less than 5% patients. Fear is intense, stark, and brief. Pleasure includes a feeling of serenity and wellbeing, though ecstatic states have been described. Anger is unprovoked, non-directive, and dissipates rapidly. The patient has no memory for his behaviour. Directive violence has **not** been shown to be of epileptic origin, in fact violent acts occur very very rarely with complex partial seizures, and are fragmentary in nature. Depression can be profound and can last for days. It frequently culminates in a generalised tonic-clonic seizure. Gelastic seizures consist of inappropriate laughter, the laugh being hollow, the facial expression unchanged, and the patient unaware later of his action. Illusions, autonomic symptoms are rare types of partial seizures.

Simple partial seizures are commonly focal motor in character, though any of the above mentioned varieties may occur, but the seizure should only be classified as simple partial if consciousness remains unimpaired throughout the seizure.

The most common type of complex partial seizure is the 'psycho-motor' variety. It consists of three phases : the first phase begins with a nonspecific aura. The aura is a feeling described by patients just prior to the onset of a seizure. It may consist of an epigastric sensation, a 'funny feeling' in the head, or feelings of unreality. Then follows a phase of total unresponsiveness, the patient being immobile. A facial grimace and head turning often herald the onset of a seizure. The head turning has no localising value, unlike that of the adverse seizure.

The second phase consists of automatisms. Automatisms in complex partial seizures may be stereotyped, consisting of aimless movements such as lip licking, lip smacking, and fumbling movements of the hands. The automatisms have no localising value, but the face and mouth are often involved and seem very susceptible to epileptiform discharge. Sometimes the patient may walk about or utter a few words which though recognisable are irrelevant to the situation. The patient is amnesic for these events. The final phase is the longest and can last upto 15 minutes. The patient is confused and gradually recovers from the seizure. Reactive automatisms occur in some patients in the postictal period.

It is important to differentiate partial complex seizures from absences. The presence of an aura, the longer duration, the complex automatisms, and the gradual return to normal consciousness help to differentiate between the two.

The diagnosis of partial seizures is sometimes very difficult and may require prolonged EEG monitoring and even the use of video recordings. A carefully taken history will suffice in all but a very small number of patients. The EEG shows localised spikes or sharp waves in an interictal recording. Use of nasopharyngeal electrodes and induction of sleep greatly enhances the likelihood of detecting the focus of abnormal discharge. Focal motor seizures in children are generally not indicative of focal brain pathology. In older patients there is a one in ten chance of finding a tumour. Other abnormalities that may be revealed after radiological tests include cysts, local or generalised atrophy, arteriovenous malformation, and infarcts.

The treatment of partial seizures is similar to that of generalised tonic clonic seizures except that valproic acid is unlikely to be of value. DPH and carbamazepine are the most useful drugs in these seizures. Patients who do not respond to medical therapy should be referred to specialised centres that undertake temporal lobectomy. Patients with unilateral anterior temporal discharge in the non-dominant hemisphere are most likely to do well after surgery. Unfortunately surgical therapy is available and indicated in only a small number of intractable patients.

Rare types of partial seizures include seizures from the supplementary motor area. The head and eyes turn to one side, the contralateral arm is raised and abducted, and the leg may be extended, during a seizure. Autonomic disturbance such as palpitations, and a flushing sensation have also been reported.

STATUS EPILEPTICUS Though previously defined as a state in which there is no recovery from consciousness between seizures, it is now defined as a condition where clinical or electrical seizure activity lasts for at least thirty minutes, whether or not consciousness is impaired. The classification of status epilepticus is given below :

TABLE 6.3

Convulsive status	Nonconvulsive status
a) Primary generalised major motor status. i. Tonic clonic status ii. Myoclonic status.	a) Spike-wave stupor or Absence status.
b) Partial or focal status. i. Focal motor status (Jacksonian Status) ii. Epilepsia partialis continua.	b) Complex partial status or psychomotor status.
c) Generalised major motor status with partial onset. i. Adversive onset ii. Partial motor onset.	

We shall restrict our discussion to major motor status as it is the most common and most serious form of status. Brief discussions of absence status and partial complex status are included so that the student is aware of these entities when he examines a case of prolonged altered consciousness.

Major motor status epilepticus 60 to 80% of cases have a focal onset, either adversive or partial motor. The clonic component of the seizure progressively decreases until only tonic seizures occur. Status epilepticus is **not** a part of the natural history of epilepsy, therefore the cause of the status must be looked for. Infection, metabolic imbalance, electrolyte disturbance, and withdrawal from alcohol are important causes of status epilepticus. Sudden discontinuation of anticonvulsant drugs is a common cause of status epilepticus. Sometimes a new neurologic condition such as the occurrence of a subdural haematoma or the presence of a neoplasm leads to status. Degenerative conditions like Lafora body disease and Kufs disease are rare causes of status.

The average duration of a status is nearly 3 hours. The mortality varies between 10 and 15%. Status epilepticus is a neurologic emergency that requires prompt attention. Permanent brain damage occurs if the status lasts for more than an hour. The Purkinje cells of the cerebellum, layers 3, 5 and 6, of the cerebral cortex, and the Sommer's sector of the hippocampus suffer the most. If adequate oxygenation is

maintained throughout the status, the degree of neuronal loss is reduced, and is restricted to the hippocampus.

The aims of therapy in status are :

1. To try and stop the status as soon as possible, preferably within thirty minutes.
2. To prevent the recurrence of seizure activity.
3. To maintain adequate oxygenation and blood pressure.
4. To correct any metabolic or electrolyte imbalances, and to search for a source of infection.

A protocol that can be followed in status is suggested below :

Maintain an airway, and if necessary intubate the patient. 50 ccs of 50% glucose may be given intravenously, *after* drawing blood for glucose, electrolytes, blood cultures if an infection is suspected, and blood for anticonvulsant drug levels if facilities for the same are available.

Start an intravenous drip, and inject 5 to 10 mgms of diazepam no faster than 2 mg per minute. The drug reaches the brain in 10 seconds and stops the seizure in nearly 70 percent of patients with status. Diazepam concentration in the brain drops drastically in twenty minutes, therefore a drug that prevents further seizures has to be given next. If intravenous DPH is available upto a gram can be given at a rate of 50 mg per minute, with close ECG and blood pressure monitoring, if monitoring facilities are not available, syrup dilantin, 2 teaspoonful (200 mgm diphenyl hydantoin) every 2 hours may be given for 5 doses, by nasogastric tube. Phenobarbital is given, in doses of 300 to 400 mg if the status is not controlled with DPH. Paraldehyde in a dose of 10 ml, 5 ml in each buttock, is very useful in many cases of intractable status. A diazepam drip, with 100 mgms of diazepam diluted in 500 ml of 5% glucose, given at a rate of 40 ml per hour is recommended in those cases that do not respond to the measures outlined above. Intravenous thiopental in a dose of 0.5 to 1 gram is given in 500 ml of normal saline, the rate of the infusion titrated to maintain the patient in a state of light anaesthesia. The thiopental drip is only recommended in the most severe cases, and anaesthesiology supervision is recommended.

NON - CONVULSIVE STATUS

Absence status It can last from hours to days, the patient having a history of absences, or generalised tonic clonic seizures. The status is precipitated by stress or trauma. The patient appears slow and has

paucity of speech. The patient may carry out daily activities such as brushing teeth, etc., but does not interact with the environment normally and appears withdrawn. Myoclonic twitches may occur. The diagnosis is made on the EEG which shows generalised spikes and slow waves. Intravenous diazepam breaks the status both clinically and electrically. High doses of Zarontin are used on a chronic basis.

Complex partial status consists of two phases that alternate in a cyclical manner, a phase of total unresponsiveness with speech arrest and stereotyped automatisms, and a phase of partial responsiveness with partial speech and quasipurposeful automatisms. Treatment is similar to that of complex partial seizures.

CHAPTER VII

PERIPHERAL NEURITIS

The peripheral nervous system consists of the anterior horn cell, its axon and descending axons of the bipolar cells of the sensory ganglion, which unite to form a peripheral nerve. Autonomic fibres join these nerves at various levels, sympathetic fibres in the cervical and thoracic region and parasympathetic fibres in the sacral regions. Peripheral neuritis encompasses any disease that affects these structures, but the term 'neuritis' is a misnomer because it implies an inflammatory process affecting the nerves. The term 'neuropathy' is considered more appropriate and diseases are described as demyelinating or axonal depending on their predominant effect on myelin or the axon. Electromyography helps in differentiating these two pathologic processes in some instances, but the previous notion that it could strictly classify the two has been discarded, since most neuropathies have both segmental demyelination and axonal degeneration.

Signs of motor involvement in peripheral nerve disease include weakness, atrophy, loss of reflexes, muscle cramps and fasciculations. Sensory features include paraesthesias, with loss of sensation in the distal parts of the extremities, the 'glove and stocking' distribution. Autonomic changes include brittle nails, a cooler limb., and a shiny dry overlying skin.

The table below lists the causes of peripheral neuritis.

TABLE 7.1

ETIOLOGIC CLASSIFICATION OF THE NEUROPATHIES

I. ACQUIRED

A. Metabolic

1. Diabetes (polyneuropathy, autonomic neuropathies, mononeuropathies, cranial neuropathies)
2. Uremia
3. Hypothyroidism (esp. carpal tunnel syndrome)
4. Acromegaly
5. Addison's disease
6. Liver failure.

B. Nutritional

1. Mixed (alcoholism, malnutrition, dietary fads, malabsorption syndrome)
2. Beriberi
3. Pellagra
4. B₁₂ deficiency
5. B₆ deficiency (pyridoxine)

C. Toxins

1. Heavy metals (lead, mercury, gold, arsenic, thallium)
2. Drugs (vincristine, INH, dapsone, nitrofurantoin, disulfiram, perhexilene, hydralazine)
3. Organophosphate compounds (tri-o-cresyl phosphate)
4. Acrylamide
5. Volatile hydrocarbons (n-hexane, methyl-n-butyl ketone)
6. Cobra venom.

D. Infections

1. Leprosy
2. Diphtheria
3. Herpes zoster
4. Botulism

E. Presumed immunologic

1. Serum sickness (esp. brachial plexus)
2. Inflammatory
 - a. motor polyradiculoneuropathy (Guillain-Barre syndrome)
 - b. sensory polyradiculoneuropathy
 - c. autonomic neuropathy
 - d. cranial polyneuropathy

F. Collagen-vascular

1. Polyarteritis nodosa
2. Rheumatoid arthritis
3. SLE
4. Scleroderma
5. Dermatomyositis-polymyositis

G. Ischemic vascular disease

H. Remote effects of cancer

1. Multiple myeloma & other dysproteinemias
2. Carcinomatous neuromyopathy

I. Trauma & compression

J. Tumours

1. Primary (neurinomas, neurofibromas, etc.)
2. Secondary

K. Miscellaneous

1. Sarcoidosis
2. Primary amyloidosis

II. GENETICALLY DETERMINED NEUROPATHIES

A. Hereditary motor sensory neuropathy (HMSN) such as

Charcot-Marie Tooth disease, Dejerine-Sottas disease, Refsum disease etc.

B. Hereditary sensory neuropathy (HSN) such as

Neuropathy of Denny-Brown, familial dysautonomia, Riley-Day syndrome congenital insensitivity to pain

C. Heredofamilial amyloidosis**D. Leukodystrophies** such as

metachromatic leukodystrophy (aryl-sulfatase deficiency)
Fabry's disease (diffuse angiokeratosis)
Krabbe's globoid cell leukodystrophy

E. Others

Acute intermittent porphyria

2. Bassen-Kornzweig disease (abetalipoproteinemia)

3. Tangier's disease (a-alphalipoproteinemia)

THE GUILLAIN-BARRE SYNDROME : ACUTE INFECTITIOUS POLYNEUROPATHY It is most commonly preceded by viral infection, though it can be associated with inoculation and mycoplasma infection. It occurs more frequently in patients with Hodgkin's disease, lymphoma and lupus. It has no age predilection, but is commonest between the ages of 20-50. It is more often seen in males than females. The symptoms begin as bodyaches, which may be accompanied by a mild fever. Some sensory complaints such as paraesthesias are common with the Guillain-Barre syndrome. An ascending motor paralysis that progresses with great speed and may leave the patient unable to move all 4 limbs, or the weakness first appears in the hip girdle and in 24 to 48 hours involve the entire lower limb.

Sensory changes are minimal in spite of the sensory complicated. The disease plateaus by 2 to 3 weeks with severe cases having upper limbs weakness and respiratory and bulbar symptoms. Respiratory muscles are involved in a quarter of patients. The patient may need a respirator for several weeks before he has recovered sufficient strength in the muscles of respiration. Bilateral or unilateral facial weakness may be present at the onset or may follow limb weakness. Optic neuritis may be rarely present. Sphincter involvement, a sensory level and signs of central nervous system involvement should lead one to question the diagnosis. Instances of autonomic dysfunction where, tachycardia and other arrhythmias, hypotension and vasomotor symptoms are present. Hypertension is present in over 30 per cent of patients. Variants of the Guillain-Barre syndrome include the Miller Fisher syndrome with its triad of ataxia, ophthalmoplegia and peripheral neuropathy.

The CSF examination shows the classical dissociation between protein and cells with protein being as high as 2 to 3 grams and cells

being less than 10. Nerve conduction velocities show slowed conductions. Presence of fibrillations and reduction of number of motor units on Electromyographic examination points to axonal involvement and is indicative of a poorer prognosis. Steroids are generally used in the treatment but their value is doubted. Prednisolone (1 to 2 mgms per Kg. body weight per day) or ACTH (40-60 units per day) are employed. The dose is soon tapered as the disease stops progressing. Plasmapheresis is presently being studied for its efficacy. The cause is not known, however, an auto-immune phenomenon is suspected that leads to demyelination of the roots with round cell infiltration. The differential diagnosis should include porphyria and botulism.

A similar picture may be seen following a number of infections such as chicken pox, typhoid, bacillary dysentery and gonococcal infection. Here the disease is less virulent and rarely affects respiratory muscles. In both instances the disease is self limiting with the majority of cases having a full recovery.

The disease considered below more often cause mononeuropathy (Single nerve involved) than polyneuropathy.

Lead Wrist drop and foot drop are common defects seen with lead neuropathy which tends to affect extensor muscles. Lead results in an exclusively motor neuropathy. Other conditions that result in exclusive motor neuropathy are porphyria, botulism and diabetic amyotrophy.

Leprosy Sensory symptoms and signs predominate. Any part of the peripheral nervous system can be affected though the ulnar and the posterior auricular nerve are more commonly involved. Leprosy leads to a mononeuritis multiplex, with different nerves being affected. Sensory loss is marked and trophic changes are seen in most cases. Rarely an asymmetrical polyneuropathy may be observed in this disease.

Herpes zoster, diabetes mellitus, periarteritis nodosa, and amyloid are other causes of mononeuropathy. The femoral nerves is the most common nerve to be affected in *diabetic neuropathy*, with the oculo-motor nerve following. Diabetic femoral neuropathy presents with pain and paraesthesias in the anterior aspect of the thigh, the pain being worse at night. Pain is severe enough to interfere with the patient's sleep. The quadriceps muscle becomes atrophic, there is weakness of leg extension with absence of the knee jerk. Relief from pain may be difficult. Analgesics, as well as Carbamazepine, and phenytoin may be

helpful, whilst control of the diabetes is imperative. Diabetes also leads to polneuropathy, having the features of a mixed sensori-motor neuropathy.

Herpes zoster virus is related to the varicella group of viruses, and causes a neuritis that occurs more commonly in elderly individuals and in patients with chronic debilitating diseases. Though it can affect any sensory nerve, it commonly tends to affect the first division of the trigeminal nerve and the intercostal nerves. The patient complains of pain and dysesthesias in the distribution of the affected nerve. A day later vesicles appear over the skin supplied by the nerve. In herpes ophthalmicus vesicles appear over the cornea. The vesicles turn into pustules and become dry in a week. In some patients pain is prominent after the vesicles dry up (post-herpetic neuralgia).

These patients suffer agonising pain that is little affected by analgesics. It is worse at night and can be triggered by touching the affected skin area. Measures to relieve pain include a trial of carbamazepine in doses of 600-1200 mgm/day, amitryptiline 30-75 mgms/day or baclofen 80-120 mgms per day and the use of transcutaneous electrical nerve stimulation. The use of high doses of steroids to relieve pain remains controversial.

Neuralgic Amyotrophy It affects the fifth and sixth cervical nerve roots and is commoner in young adults affecting males more often than females. Beginning with excruciating pain in the shoulder region, there is rapid development of muscle wasting and weakness. There is atrophy in the muscles supplied by the fifth and sixth cervical roots, with other nerve roots being only rarely involved. Pain disappears in a few days leaving muscle weakness and wasting, only a few patients do not recover completely from the muscle paralysis. The opposite side may be involved simultaneously or 1 to 3 months following the initial attack. A recurrent familial form has also been described. This disease is also known as the Parsonage-Turner syndrome and can occur after injection of sera. Brachial plexus lesions are also seen with direct infiltration of tumour as in carcinoma of the lung, after trauma, and rarely after radiation.

Vitamin deficiencies are a very common cause of neuropathy in our country and result from deficiency of B₁, and nicotinic acid. Patients who suffer from chronic disease of the colon, chronic alcoholics and malnourished individuals may show pallor, cheilosis around the angle of the mouth, a red raw tongue, and hyperkeratosis. Signs of vitamin A and

D deficiency may be present simultaneously. In pellagra, dementia and diarrhoea accompany the peripheral neuropathy. Paraesthesias are common, there being distal sensory loss, and absence of deep tendon reflexes. Nerve conduction may be slowed and EMG may show signs of denervation.

Pernicious anaemia is very rare in India, but vitamin B₁₂ deficiency resulting from a malabsorption syndrome, does occur. Vitamin B₁₂ deficiency generally present along with other vitamin deficiencies. Wernicke's encephalopathy occurs in chronic alcoholics and in malnourished patients. It consists of ataxia, ophthalmoplegia, nystagmus, and peripheral neuropathy. There may be problems with memory. Treatment with intravenous thiamine in doses of 100 to 500 mgms will result in remission of symptoms in the majority of patients.

HEREDITARY SENSORI-MOTOR NEUROPATHY :

These may represent as many as 25% of all patients with an undiagnosed peripheral neuropathy. 4 prominent varieties are described below :

Peroneal muscular atrophy (Charcot Marie Tooth Disease) It is an autosomal recessive disease though it may occasionally be seen as a sporadic case. Symptoms begin in adolescence and are commoner in male members of the family. There is a bilateral foot drop, with wasting that tends to involve the small muscles of the feet, producing pes cavus. The anterior compartment of the leg is next to undergo atrophic changes, with the wasting ascending proximally to produce the classical 'stork leg' or 'inverted champagne bottle' abnormality. Motor features predominate, though loss of vibratory sense may be found in the ankles. The ankle jerks and at times the knee jerks are absent. A few years later wasting has involved the distal parts of the upper extremity, with minimal sensory changes. Nerve conduction velocities are markedly slowed. The disease does not shorten the life span but does cause considerable disability.

Dejerine Sottas type of hereditary sensori-motor neuropathy This is an autosomal dominant disease that manifests itself in infancy and early childhood. Signs of a sensori-motor neuropathy are detected in all four limbs. The condition is slowly progressive, with thickening of the nerves. In India these cases may be mistaken for leprosy, another cause of thickened nerves. There may be cranial nerve involvement in some

cases. Motor milestones are not achieved and incapacitation, with the patient leading a bedridden existence can be seen in this disease.

Refsum's hereditary ataxic neuropathy It is due to a disorder of metabolism which leads to increased levels of phytanic acid. This disease is very rare in India. Ichthyosis, deafness and retinitis pigmentosa accompany the peripheral neuropathy.

HEREDITARY SENSORY NEUROPATHIES

These usually present in childhood, with the occurrence of non-healing trophic ulcers, that are unaccompanied by pain. Total loss of pain and other modalities of sensation is observed. Multiple trauma to joints that are lacking the sensation of pain, leads to a destructive arthropathy (Charcot's joint).

The underlying lesion is usually in the dorsal root ganglia.

Other forms of hereditary neuropathy like the amyloid and lipoprotein deficiencies are very rarely seen in India.

We shall now discuss some mononeuropathies that result from a nerve being entrapped under a bony or fibrous structure.

CARPAL TUNNEL SYNDROME This is the commonest entrapment syndrome and is usually seen in obese females. The median nerve is entrapped as it traverses under the flexor retinaculum at the wrist. Bilateral lesions are not uncommon. The patient complains of pain and paraesthesia that are worse at night. In severe cases there is weakness and wasting of the thenar muscles. The symptoms involve the thumb, the index and middle fingers, but never the little finger. The symptoms progress and may be accompanied by detectable sensory loss. In early cases the Phalen's sign may be the only positive sign on examination. It consists of pain in the median distribution on extreme flexion of the wrist for 1-2 minutes. Tinel's sign may also be positive but not considered as diagnostic of this syndrome. Tinel's sign consists of paraesthesias that result after percussing over the nerve. The diagnosis is made by the increase in distal latency (normal 4.5 milliseconds for motor and 3.5 milliseconds for sensory) on Nerve Conduction. Severe cases may show slowing of median nerve conduction velocity due to retrograde degeneration along the nerve. Causes include acromegaly, rheumatoid arthritis and myxoedema. Treatment is surgical decompression which afford immediate relief. Local injection of steroids may provide symptomatic relief in early cases.

THORACIC OUTLET SYNDROME

It is the next most common cause of nerve entrapment. It can occur due to a cervical rib or due to a fibrous band that compresses the lower roots of the brachial plexus (C 7,8, T 1) and the subclavian artery. Pain generally occurs at night due to relaxation of the elevators of the shoulder. Pain occurs along the median border of the forearm and the ulnar side of the hand. The pain is shooting and burning in character. Raynaud's phenomenon is observed on exposure to cold or on immersing the hand in cold water. Examination may reveal decreased sensation along the C7,8, T1, dermatomes. The radial pulse is feeble and bruit may be heard over the supraclavicular fossa. The Addson's test may be positive. It consists of asking the patient to raise his arm at the shoulder and at the same time brace the shoulder backwards, turn the head to the same side and take a deep breath. The radial pulse is obliterated and symptoms are reproduced if the test is positive. X-ray of the thoracic outlet region may show either a cervical rib or a beaked and enlarged transverse process of the seventh cervical vertebra. Treatment consists of excision of the cervical rib, or of the fibrous band. Some cases with mild symptoms may benefit by shoulder exercises alone.

Meralgia paraesthetica Though common in obese females it has been observed in very thin people. The lateral cutaneous nerve of the thigh is entrapped in the inguinal ligament with the patient complaining of paraesthesias in the lateral aspect of the thigh mainly on walking. Examination reveals an oval area of anaesthesia in this region. Trophic changes are not seen. Steroid injected in the inguinal ligament around the nerve may provide relief for most patients, though in some cases surgical intervention to free the nerve may be necessary.

Other nerve entrapment syndromes include the tarsal tunnel syndrome, the cubital tunnel syndrome, and the anterior interosseous nerve syndrome. These syndromes are relatively rare and are therefore not described here in detail.

CERVICAL AND LUMBAR SPONDYLOSIS

Prolapse of the intervertebral disc results in compression of nerve roots and causes radicular pain. Disc prolapse involves the cervical and lumbar vertebral regions frequently, though rarely the thoracic region may be affected. Intervertebral discs prolapse dorsally because the anterior longitudinal ligament is very strong and does not give to allow

anterior disc protrusion. Acute disc prolapse generally follows minor trauma, and in the cervical region is secondary to a 'whiplash'. 'Whiplash' occurs when the neck is subject to sudden jerking as in the sudden application of brakes to a fast moving vehicle. Lumbar disc prolapse usually occurs on lifting heavy objects. In India a common cause of an acute lumbar disc prolapse is the lifting of a bucket filled with water. The nucleus pulposus prolapses through a tear in the annulus fibrosus and forms a mass that usually impinges laterally to compress a nerve root in the intervertebral foramen. If the prolapse occurs medially it will compress the spinal cord in the cervical region and the cauda equina in the lumbar area. Nerve roots can also be entrapped in the foramina due to spondylotic changes and formation of osteophytes.

Cervical roots C5 and C6 are most commonly involved in disc disease. An acute disc prolapse leads to shooting pain along the nerve that is compressed. A C5 lesion causes pain in the shoulder and a C6 root involves the arm. There is neck stiffness and pain in the nuchal region with spasm of the extensor spinae muscles. These muscles are supplied by the anterior division of the involved nerve roots. Movements of the neck cause severe pain. There is sensory loss over the deltoid (C5) and sometimes numbness of the thumb. The patient is unable to abduct the shoulder against resistance. If only the C5 root is involved tendon reflexes are normal. C6 involvement leads to loss of the biceps and supinator jerks and to weakness of elbow and wrist flexion. There may be sensory loss over the outer border of the forearm. If there is simultaneous compression of the cervical spinal cord an inverted supinator jerk may be elicited. Here, absence of the supinator jerk is associated with finger flexion due to an overflow phenomenon.

Chronic cervical disc disease (Spondylosis) is accompanied by moderate pain and may cause long tract signs if a myelopathy has also occurred. Sensory changes are minimal (peri-anal region not spared) but pyramidal signs are present. Sphincter involvement is rare, the patient complaining of urgency and hesitancy of micturition and constipation.

Lumbar disc prolapse commonly involves the L5 S1 interspace. A high lumbar disc lesion is very rare and if suspected, the student must first carefully rule out either a femoral neuropathy or plexus involvement. Severe spasms of the extensor spinae muscles cause pain in the lower back and restrict movements of the lumbar spine. Pain may radiate along the

sciatic nerve with the patient complaining of pain that goes down the back of the thigh and leg. Paraesthesias occur along the lateral aspect of the leg and the medial border of the foot. On examination the straight leg raising (SLR) test is positive. Stretching of the sciatic nerve as the leg is flexed at the hip causes pain along the back of the leg. This test is also known as Lasegue's sign. A sensory loss may be detected in the L5 S1 dermatomes. Weakness of dorsiflexion of the great toe is seen in most cases. In a massive disc prolapse foot drop may occur, with rapid development of sphincter incontinence. Fortunately these cases are rare. Loss of ankle jerk and of the medial hamstring reflex is noted. EMG shows absence of the H reflex on the side of the lesion. The H reflex is a late response observed on submaxial stimulation of the posterior tibial nerve, and is affected by lesions that involve the nerve proximally. Fibrillations may be seen in the affected muscles and in the paraspinal region. Signs of increased muscle irritability such as an increase in insertional activity and positive sharp waves are frequently encountered in acute disc prolapse.

Treatment for acute disc prolapse consists of strict bed rest, analgesics and gradual exercises to relax and then strengthen muscles. Cervical collar and lumbar braces may provide symptomatic relief. Traction is of uncertain value. Surgery is called for when pain continues in spite of vigorous conservative measures. Whenever there is any evidence of cord compression surgery must be considered immediately. Lumbar disc prolapse that causes sphincter problems should also be treated surgically.

A patient with a congenitally narrow spinal canal may have symptoms of cord compression with minimal degenerative changes. In the lumbar region a condition known as lumbar canal stenosis develops with symptoms of intermittent claudication. The patient complains of pain along the nerve roots when he walks a short distance. The pain is relieved by rest. Examination reveals a normal arterial system but signs of root involvement may be detected.

Myelography should be done in patients with definite signs of disc disease that results in neurologic deficit. If EMG facilities are available an EMG should be done prior to myelography. However the EMG may be negative if done too early in the course of the disease.

CHAPTER - VIII

DISORDERS OF MUSCLE

The word muscle comes from the Latin word *Musculus*—meaning a mouse, as the shape of muscle appeared to resemble a mouse. Muscle is the largest organ of the body and forms a fifth of the body weight. There are about 600 different muscles in the body.

A muscle fibre is an elongated cylindrical multinucleated cell. The length of the fibre varies from a few millimeters to many centimeters and the diameter of each cell varies from 10 to 100 microns. The fibre is bound by Sarcolemma. The motor end plate is a specialised region of the muscles where a motor nerve makes functional contact with the muscle. Muscle fibre contains fine fibrillar structures called filaments which are made of actin and myosin. These are arranged in a highly ordered fashion and are the contractile system of the muscle. The sarcoplasmic reticulum and the 'T' system are the tubular networks which play an active part in the contraction and relaxation of the muscle fibre. The cytoplasm (sarcoplasm) also contains mitochondria, golgi apparatus, glycogen granules, lipid droplets and ribosomes.

The alpha motor neurones supply the extrafugal muscle fibres. Each motor neurone supplies 15 to 200 muscle fibres depending on the size and function of the muscle. The alpha motor neurone with its axon and the number of muscle fibre supplied by it, form the physiological unit called 'motor unit'.

In 1968 the World Federation of Neurology classified disorders of voluntary muscle. A simplified version of the same is given below.

Myopathy, has been defined as any, physiological, structural, biochemical or electrical alteration of muscle fibre and its connective tissue without the involvement of the peripheral or central nervous system. The word dystrophy should be only used for genetically inherited primary disorders of muscle fibres. Hence, all dystrophies are myopathies but all myopathies are not dystrophies.

TABLE 8.1

CLASSIFICATION

**A. GENETICALLY DETERMINED MYOPATHIES
(DYSTROPHIES)**

- | | |
|-------------------------------|---|
| I a) Sex-linked | <ul style="list-style-type: none"> i Severe-Duchenne variety iii Mild - Becker variety |
| b) Autosomal recessive | <ul style="list-style-type: none"> i Autosomal recessive dystrophy of childhood ii Limb - girdle (could be sporadic) iii Myopathy limited to Quadriceps iv Ocular (could be sporadic) v Oculo-pharyngeal |
| c) Autosomal dominant | <ul style="list-style-type: none"> i Facio-scapulo-humeral ii Distal myopathy iii Scapulo-peroneal variety. |

II Congenital myopathies

- a) Congenital muscular dystrophy
- b) Benign congenital hypotonia
- c) Central core disease
- d) Nemaline or rod body myopathy
- e) Mitochondrial myopathy
- f) Centronuclear myopathy
- g) Congenital fibre type disproportion.

III Myotonic disorders

- a) Dystrophia myotonica
- b) Myotonia congenita
- c) Paramyotonia

IV Storage Disorders

- a) Glycogen storage
- b) Lipid storage.

B. TRAUMA**I Physical**

- a) Crush syndrome
- b) Anterior or posterior tibial compartment syndrome.

II Toxic — Snake bite, Alcohol.**III Drugs — Steroids, Chloroquine, Emetine, Vincristine, Clofibrate, Amphotericin, Polymixin, Triorthocresyl phosphate,****C. INFLAMMATORY**

- a) Viral**
 - i Bornholm disease
 - ii Influenza
 - iii Coxsackie virus
- b) Bacterial**
 - i Gas gangrene
 - ii Tetanus
 - iii Syphilitic myositis
- c) Parasitic**
 - i Cysticercosis
 - ii Trichinosis
 - iii Toxoplasmosis
- d) Auto-immune**
 - i Polymyositis Acute, Subacute, and Chronic
 - ii Dermatomyositis – Associated with SLE, scleroderma
 - iii Polymyositis associated with Malignant disease
 - iv Polymyositis associated with Sarcoid, Giant cell arteritis polymyalgia rheumatica.

D. ENDOCRINAL MYOPATHIES AND METABOLIC MYOPATHIES

- a) Thyroid**
 - i Thyrotoxicosis - Myopathy, Myotonia and Periodic paralysis.
 - ii Hypothyroid - Myopathy, Debre-Semlaigne-Kocher syndrome Hoffman's syndrome.

b) Pituitary

- i Acromegaly
- ii Cushing's disease
- iii Exophthalmic ophthalmoplegia
- iv Hypopituitary
- v ACTH Myopathy

c) Adrenal

- i Addison's disease

d) Parathyroid

Hyperparathyroidism

e) Metabolic

- i Osteomalacic- Malnutrition, Steatorrhea
Malabsorption Renal
acidosis.
- ii Nutritional -Protein calory malnutrition

Alcoholic.

E OTHER MUSCLE DISEASES

- a) Carcinomatous neuromyopathy
- b) Paroxysmal myoglobinuria
- c) Muscle Hypertrophy
- d) Disuse atrophy
- e) Tumours of the muscle.

DUCHENNE TYPE OF GENETICALLY INHERITED MYOPATHY

The commonest dystrophy met with in clinical practice, -Duchenne's dystrophy-is a sex-linked disorder only affecting males. The disease is recognised by the parents by the third or fourth year of the child's life and from then on progresses relentlessly.

The child is normal at birth but may have delayed motor milestone. Some children walk normally till the age of 3, whilst others have difficulty with gait from the very beginning. The parents notice that the child waddles, falls frequently when walking, and cannot run. Hypertrophy of the calves may be obvious to observant parents. The child soon has difficulty climbing stairs and is unable to raise himself from the

floor without support. He uses a characteristic manoeuvre (Gower's sign) to assume the upright position. He first goes down on all four limbs, swings the arms to the knees, then climbs up his legs as it were, and stands upright. When erect, he stands with his feet apart, his abdomen protrudes and the lumbar lordosis is increased. Weakness of the arms is noted by the family two to three years after the onset of the disease, though it can be demonstrated much earlier on physical examination.

The weakness progresses to involve all four limbs, the muscles of the neck and trunk, the child walking with a wide base and a marked lumbar lordosis—the waddling gait of muscular dystrophy. Tightening of the tendo-achilles causes the patient to walk on his toes. The limbs become markedly weak, the child is confined to a wheelchair, and contractures develop in the hamstrings, and at the hips and elbows. Unless great care is taken spinal deformities like scoliosis occur. By the time the child reaches the age of 16 he is confined to bed, death occurs around 20 to 25 years of age secondary to respiratory infection or cardiac failure.

On physical examination the child appears mildly retarded with an I.Q. around 70. There is wasting and weakness of the limb muscles, with the iliopsoas, the quadriceps, and the glutii amongst the first to be affected. Weakness then spreads to involve the tibialis anterior and the peronei. In the arms, the pectorals, the latissimus dorsi, deltoids, triceps, biceps, and brachioradialis are involved early in the course of the disease. Winging of the scapula is very prominent in the late stages. Power is better maintained in the hamstrings compared to the quadriceps, the gastrocnemius compared to the tibialis anterior, in the flexors of the wrist compared to the extensors, and in the extensors of the neck compared to the flexors. In the late stages the intercostal and abdominal muscles become weak.

Hypertrophy of the calves is seen in almost all cases, and of the tongue in 20 to 25% of cases. Hypertrophy of the deltoids, the flexors of the forearm, the supraspinatus, infraspinatus, and the gluteal muscles may also be seen. Rarely the masseters may show hypertrophy. The tendon reflexes are decreased and then lost in the upper limbs early in the course of the disease, the knee jerks are next to disappear but the ankle jerks are preserved till very late. This is due to sparing of the gastro-soleus till late in the disease. The electrocardiogram is abnormal in 60 to 80% of cases, showing a labile tachycardia and a variety of disturbances of rhythm. Serum enzyme (CPK, SGOT, SGPT, LDH and Aldolase) levels are elevated, with the CPK values reaching 20,000 units in the early stages, and falling as the disease progresses.

Muscle biopsy shows loss of fascicular pattern, with variation in fibre size. Hyalinization, flocculo-granular change and necrosis occur. Regeneration of muscle fibres is a prominent feature. Nuclear migration and pyknotic changes are seen. There is a marked increase in connective tissue and in adipose tissue. There is no known therapy, but physiotherapy may help to prevent formation of contractures and deformities. Tests that detect carriers of the gene of muscular dystrophy, include enzymatic levels, study of ribosomes, EMG and muscle biopsy. The sex of the child is determined at twelve weeks in a pregnant carrier woman. If the child is a male, termination of pregnancy is advised. Measures such as this are the only way to prevent the disease at present.

BECKER TYPE The disease begins between the ages of 5 to 15, the symptoms are similar to Duchenne's with hypertrophy of the calves being present in all cases. The disease progresses slowly as compared to Duchenne's dystrophy, the patient being able to walk till he is in his mid twenties, and there is often no shortening of the life span. Cardiac involvement is rare, as are contractures. The detection of the carrier state is less satisfactory than in Duchenne's disease. Treatment consists of physiotherapy.

LIMB GIRDLE DYSTROPHY Inherited as an autosomal recessive disorder it can occur in either sex. Symptoms of pelvic girdle weakness appear around the second or third decade, with muscles of the shoulder girdle becoming involved later in the disease. Life expectancy is shortened to some extent, with the patient confined to bed in the fourth to fifth decade of life. Cardiac involvement is extremely rare.

AUTOSOMAL RECESSIVE DYSTROPHY OF CHILDHOOD Symptoms appear by adolescence and are similar to those seen in Duchenne's dystrophy. Slight asymmetry of signs may be seen, the course is slow, with survival upto the third or fourth decade being common. The ECG is normal, and the CPK though high does not reach the massive levels of Duchenne's dystrophy. Treatment is similar to other dystrophies.

CONGENITAL MUSCULAR DYSTROPHY This type of dystrophy is more common in India than the two described above. The inheritance is of the autosomal recessive type. The child is hypotonic at birth. There are two types : the severe and the benign variety. The severe variety progresses faster than Duchenne's disease, contractures appear early, the life span is markedly shortened with the majority dying into their teens. The benign variety progresses very slowly, the life span is normal,

and the weakness minimal. Contractures hardly ever occur, the CPK is minimally elevated or may even be normal, the ECG shows no abnormality and the diagnosis is made on muscle biopsy. Sections of muscle show dystrophic changes associated with excessive collagen formation and minimal regeneration.

FACIO—SCAPULO—HUMERAL DYSTROPHY It has a dominant inheritance, with an onset that varies from childhood to adult life. In some patients the weakness is so minimal that they go through life without any symptoms. The facial and shoulder girdle muscles are the first to be involved, there is winging of the scapula and an inability to raise the arm above the shoulder. The muscles of the pelvic girdle are affected late. Muscle hypertrophy is rare, the disease has a benign course with no shortening of the life span. In very few patients the course may be rapid, and incapacitation occurs early in life.

OCULAR MYOPATHY Ocular muscle involvement is not seen in any of the dystrophies described above. The disease appears at any age, though generally the onset occurs in the first or second decade. In India the majority of patients have no family history. The first features are a bilateral symmetrical ptosis that shows no diurnal variation. Bilateral symmetrical weakness of extra ocular muscles follows, leading to limitation of eye movements in all directions. There is no diplopia because the weakness is symmetrical. The weakness may progress to involve the pharyngeal and facial muscles in some patients.

MYOTONIA CONGENITA The symptoms appear in the first decade, though myotonia may be present from birth. Myotonia is wide spread in the body musculature and is worsened by cold weather. The myotonia results in difficulty in getting up and may produce frequent falls. Some patients may have the presenting complaint of difficulty in relaxing their grip. The ECG shows the classical high frequency discharges, the sound of which has been compared to that of a divebomber. Quinine, procainamide, or phenytion help to relieve the myotonia, which diminishes gradually with age.

MYOTONIA DYSTROPHICA The onset is around 20 to 30 years of age. The facial appearance is characteristic, a mild ptosis, lack of facial expression, and wasting of the masseters, and temporalis muscles is prominent. Frontal baldness is common. Distal limb weakness with difficulty in relaxing the grip is noted. Weakness may spread to involve the proximal musculature. If the tongue muscles are involved, dysarthria

occurs. A variety of other abnormalities occur that include cataract formation, cardiac involvement, testicular atrophy, and endocrine dysfunction namely diabetes. The patient is gradually crippled by the disease and is confined to bed about 15 to 20 years after the onset of symptoms. There is no treatment for the weakness, but the myotonia can be relieved by the use of phenytoin or procainamide.

MYASTHENIA GRAVIS Though this is not a true disorder of muscle in the strict sense of the word, it is included in this chapter for the sake of convenience. The disease is known worldwide, can occur at any age, though peak occurrence is around the second and third decades. It is commoner in females before the age of thirty and in males after the age of forty. The disease is divided into four sub types based on its responsiveness to therapy :

- i) Ocular myasthenia
- ii) A - Mild generalised myasthenia responsive to drugs.
B - Moderate generalised myasthenia unsatisfactory response to drugs
- iii) Acute fulminant myasthenia, patients present in crises, thymoma more common.
- iv) Severe myasthenia with a high mortality, similar to type III but occurring after a few years of type I or II.

Myasthenia is associated with thyrotoxicosis in 5% of cases, and is often associated with immunological disorders such as systemic lupus erythematosus, rheumatoid arthritis, and pemphigus.

Acetyl choline is synthesized at the terminal end of the motor axon and stored in vesicles. A single nerve action potential triggers a release from 100 to 200 vesicles, acetyl choline is now available for binding to receptors in the myoneural junction of the muscle membrane. Increased permeability of membrane to sodium, potassium and calcium allows an influx of ions that start the wave of depolarisation and convert chemical energy to electrical energy.

The isolation of antibodies to acetyl choline receptors in many patients with myasthenia, and the production of experimental autoimmune myasthenia in laboratory animals, may indicate an autoimmune pathogenesis for myasthenia. Presence of acetyl choline receptor antibody in the serum provides a sensitive test for the diagnosis of myasthenia, but levels of antibody do not correlate with either activity or severity of the disease. The role of the thymus and cellular immunity is

still unclear. One hypothesis argues that immunologically competent 'T' cells, infiltrate the neuromuscular junction and destroy the acetyl choline receptors, co-operating with the helper 'B' cells to form antibodies against these acetyl choline receptors.

Clinical features The most important clinical feature of myasthenia is easy fatiguability of muscles that is worse on exercise and relieved by rest. Symptoms have a diurnal variation, with the patient feeling worse in the evening. The muscles supplied by the cranial nerves are often involved, with resultant diplopia, ptosis, ophthalmoplegia, dysphagia, dysphonia. The disease then involves the proximal limb muscles. Rarely, weakness appears in the limbs before it appears in the cranial muscles. Muscle involvement is usually asymmetric, with muscles of the neck, shoulder girdle, and the flexors of the hip being more frequently involved than muscles of the trunk and legs. Drooping of the eye-lids (ptosis) is the commonest symptom. The disease begins insidiously, but may present acutely after severe emotional stress. Myasthenic patients feel better in cold weather compared to warm weather. Exposure to bright sunlight affects the patient adversely.

Physical Signs The signs associated with myasthenia vary from patient to patient, and depend upon the muscle groups that are involved. Ptosis may improve after eye closure for a minute or may worsen if the patient is asked to look up continuously for about a minute. The deep tendon reflexes are brisk, and there is no atrophy till late in the course of the disease.

Diagnosis The diagnosis can be confirmed by the use of electrophysiological and pharmacological tests. Repetitive stimulation of a motor nerve results in a progressive decrease in the amplitude of the evoked potential. Cooling the area reduces the amount of decrement produced after repetitive stimulation.

Edrophonium hydrochloride (Tensilon) given in a dose of 10 mgm intravenously produces rapid improvement of signs. The response is dramatic and confirms the diagnosis of myasthenia. If Tensilon is not available, Neostigmine is given in a dose of 0.04 mgm per Kg. body weight subcutaneously and the patient observed for 15 to 30 minutes. The muscarinic effects of these drugs can be blocked by giving atropine 0.4 mg intramuscularly prior to the injection of either Tensilon or Neostigmine.

Management The treatment of a myasthenic has to be individualised to meet the needs of a particular patient. In patients with ocular myasthenia alone, drugs that improve neuromuscular transmission, are sufficient to provide relief. Neostigmine in a dose of 15 mgm every 4 to 6 hours, or pyridostigmine 60 mgm three times a day, are generally all that is required. Pyridostigmine is a longer acting drug.

The treatment of other types of myasthenia should include a consideration of thymectomy at any early stage. Removal of the thymus may provide a cure especially in those patients who do not have a thymoma. Plasmapheresis is the treatment of choice for a myasthenic crises. A 'myasthenic crises' is one in which the patient becomes refractory to drugs and has severe respiratory distress. The patients should be placed on the respirator and all drugs discontinued for a short time. Myasthenic patients may also suffer from a 'cholinergic crises' that results from excessive anticholinesterase medication. The Tensilon test is used to differentiate between the two types of crises. The patient with a myasthenic crises becomes stronger after an injection of Tensilon, whereas a patient with cholinergic crises becomes weaker after the Tensilon is injected. Sometimes the differentiation is not clear cut, and it is advisable to put the patient on a respirator and discontinue all drugs. Steroids are very helpful in the treatment of moderate to severe cases of myasthenia. Prednisone in doses of 60 to 80 mgm a day is given daily initially and then gradually reduced to a dose of 20 to 40 mgm on alternate days. Whenever steroid therapy is begun, it is necessary to hospitalise the patient as some patients become very weak in the first few days of steroid therapy. More recently immunosuppressive drugs are being used, in refractory myasthenics, with some success.

Spinal muscular atrophy Though this condition is due to primary degeneration of the anterior horn cells of the spinal cord, it is included in this chapter as it presents and behaves very much like a muscular dystrophy, and therefore may cause some confusion in the diagnosis, of the dystrophies.

The spinal muscular atrophies are a group of inherited disorders with a wide spectrum of clinical features. The pattern of inheritance is autosomal recessive, though dominant and sex-linked forms have been described.

Juvenile spinal muscular atrophy (Wolfhart - Kugelberg - Welander syndrome) The earlier symptoms consist of difficulty in rising from the

squatting position, and in climbing up the stairs. The patient falls frequently and has an abnormal gait that resembles the gait of muscular dystrophy. The disease starts in the first decade of life. The majority of patients have a weakness of both the upper and lower limbs. About half the patients have weakness of the trunk and neck muscles. The hamstrings are involved to a greater extent than the quadriceps, the exact opposite of the predominance of weakness in dystrophy. Affected muscles are hypotonic and atrophic, with the atrophy being most pronounced proximally. Fasciculations are not seen frequently, but when present confirm the diagnosis of spinal muscular atrophy. The tendon reflexes are absent in most cases, but some patients may have preservation of ankle jerks, as is often seen with the dystrophies.

If the disease begins early in life the course is likely to be rapid, with the patient confined to a wheelchair by the second decade of life. The later the onset of symptoms the milder the clinical deterioration and the slower the course. The serum enzymes (CPK, SGOT, SGPT, LDH) may be elevated, the ECG is normal, and the EMG shows features of a neurogenic disorder. Muscle biopsy and histochemistry are diagnostic of the disease. Treatment consists of supportive physiotherapy and other rehabilitative measures.

CHAPTER - IX

INTRACRANIAL TUMOURS.

Intracranial tumours can be **primary**, arising from neuroectodermal tissue, the meninges, or **secondary**, invading the cranium or intracranial structures due to direct or haematogenous spread. The cause of intracranial tumours is as yet unknown though certain viruses like the myxoma virus and chemicals such as tar have been shown to be carcinogenic in animals.

Intracranial tumours can be classified according to their location, cell of origin, or growth characteristics. We find the classification used by K.J. Zulch useful and therefore print it in detail below :

TABLE 9.1

CLASSIFICATION OF INTRACRANIAL TUMOURS

(modified from the International Histological
Classification of Tumours)

1. TUMOURS OF NEUROEPITHELIAL TISSUE
 - A. **Astrocytic Tumours** Astrocytoma, Astroblastoma, Malignant astrocytoma.
 - B. **Oligodendroglial Tumours** Oligodendroglioma, Malignant oligodendroglioma.
 - C. **Ependymal and Choroid plexus Tumours** Ependymoma, Malignant ependymoma, Choroid plexus papilloma, malignant papilloma.
 - D. **Pineal Cell Tumours** Pinealocytoma, Pinealoblastoma.
 - E. **Neuronal Tumours** Gangliocytoma, Ganglioglioma, Ganglioneuroblastoma, Neuroblastoma.
 - F. **Poorly Differentiated and Embryonal Tumours** Glioblastoma, Medulloblastoma, Spongioblastoma, Gliomatosis cerebri.
2. TUMOURS OF NERVE SHEATH CELLS
 - A. **Neurilemmoma** (Schwannoma, Neurinoma)
 - B. **Malignant Neurilemmoma**
 - C. **Neurofibroma**
 - D. **Malignant Neurofibroma**

3. TUMOURS OF MENINGEAL AND RELATED TISSUE.
 - A. **Meningioma.**
 - B. **Meningeal Sarcomas**
 - C. **Xanthomatous Tumours** Fibroxanthoma, Xanthosarcoma.
 - D. **Primary Melanotic Tumours** Melanoma, Meningeal Melanomatosis.
4. PRIMARY MALIGNANT LYMPHOMAS
5. TUMOURS OF BLOOD VESSEL ORIGIN
 - A. **Haemangioblastoma.**
6. GERM CELL TUMOURS.
 - A. **Germinoma**
 - B. **Embryonal Carcinoma**
 - C. **Choriocarcinoma**
 - D. **Teratoma**
7. OTHER MALFORMATIVE TUMOURS AND TUMOUR-LIKE LESIONS.
 - A. **Craniopharyngioma**
 - B. **Rathke's Cleft Cyst**
 - C. **Epidermoid Cyst**
 - D. **Dermoid Cyst**
 - E. **Colloid Cyst of Third Ventricle**
 - F. **Enterogenous Cyst**
 - G. **Other Cysts**
 - H. **Lipoma**
8. VASCULAR MALFORMATIONS
 - A. **Capillary Telangiectasia**
 - B. **Cavernous Angioma**
 - C. **Arteriovenous Malformation**
 - D. **Venous Malformation**
 - E. **Sturge – Weber Disease** (Cerebrotrigeminal Angiomatosis)
9. TUMOURS OF THE ANTERIOR PITUITARY.
 - A. **Pituitary adenoma** Acidophil, Basophil, Mixed and Chromophobe
 - B. **Pituitary Adenocarcinoma**

10. LOCAL EXTENSION FROM REGIONAL TUMOURS

- A. **Glomus Jugulare Tumour** (Chemodectoma, Paranganglioma)
- B. **Chordoma**
- C. **Chondroma**
- D. **Chondrosarcoma**
- E. **Olfactory Neuroblastoma**
- F. **Adenoid Cystic Carcinoma** (Cylindroma)

11. METASTATIC TUMOURS

12. UNCLASSIFIED TUMOURS

Infective granulomas like tuberculomas, abscesses, and parasitic cysts should not be considered as tumours of the central nervous system, even though they produce symptoms and signs that are kin to tumours. Tumours may behave as space occupying lesions, they may compress important structures, they may destroy tissue, or destruction and growth occurs paripassu as in microgliomas and gliomatosis cerebri.

Clinical Features The clinical features of an intracranial mass depend on its location, its rate of growth, and its ability to infiltrate tissue. Cerebral oedema develops with most fast growing tumours and causes further increase in neurologic deficit. As the brain is encased in the tight bony compartment of the skull, the tumour does not have much scope for enlargement and before long it causes changes in the sensorium and produces signs of increased intracranial pressure. Some tumours may bleed into their substance causing sudden increase in signs and appearing like a vascular event. Glioblastoma multiforme and oligodendroglioma are most often associated with such an event. The intracranial cavity is further divided into infra and supratentorial compartments, and tumours in either compartment displace structures and lead to signs of herniation (Fig 9.1)

Intracranial pressure is the pressure under which CSF exists in the cranial cavity. It can be measured by the introduction of a catheter into the lateral ventricle and recording the pressure on an electronic recording apparatus. Modern techniques monitor intracranial pressure continually by the use of transducers that are placed subdurally. Normal intracranial pressure varies between 150-200 mms of CSF. Remember that the brain, the CSF and the blood that flows through this region (normally a fifth of cardiac output), are all contained in a rigid bony case—the skull—with the foramen magnum providing a small vent for

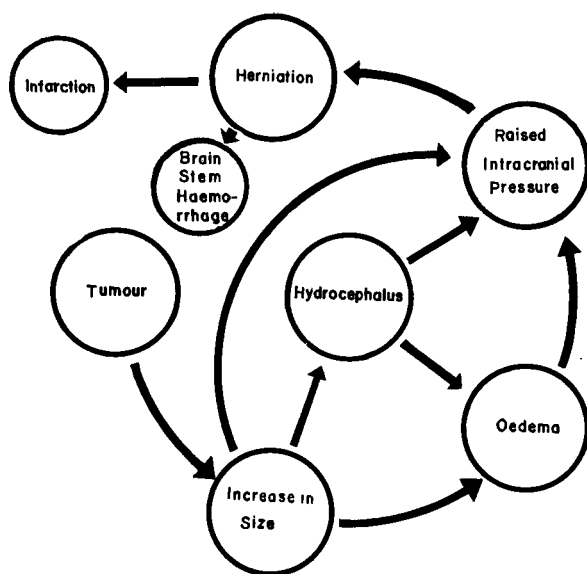


Fig. 9.1 : Flow diagram showing progressive complications of an intracranial tumour

overflow of CSF. Therefore an increase in any one of the three components (brain, CSF, blood) must result in displacement of the other two. Small and gradual rises in intracranial pressure can be accommodated without significant effects, but sudden and large increases in pressure are dangerous and can prove fatal if remedial measures are not taken on an emergency basis. These effects are due to the result of brainstem dysfunction that accompanies the rostro-caudal deterioration. Depending on the site of the lesion responsible for these events, first the cingulate herniates under the falx, then the uncus of the temporal lobe herniates under the tentorium and finally the medulla herniates through the foramen magnum (Fig 9.2). Increases in CSF pressure alone does not lead to herniation, as in pseudotumour cerebri, but any event that causes obstruction to flow of arterial blood or spinal fluid begins the process. Arterial blood continues to be pumped in, accelerating the herniation, till intracranial pressure rises above the level of arterial pressure. When this occurs there is no intracranial blood flow, a sign that can be demonstrated on angiography and constitutes one of the tests for documenting 'brain death'.

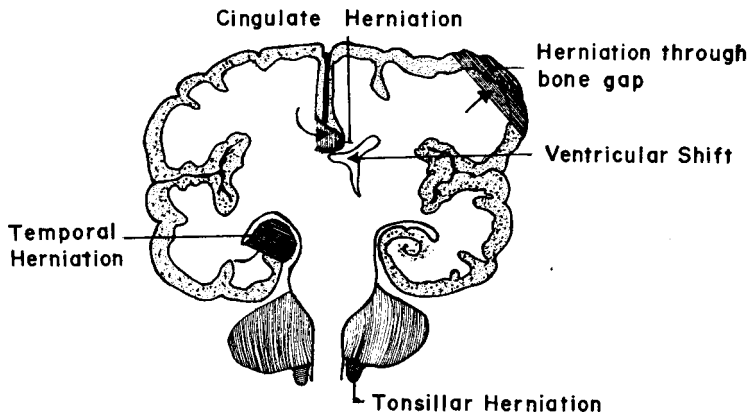


Fig. 9.2 : Coronal section of the brain showing different types of herniations

Causes of increased intracranial pressure are legion. Besides tumours they include a) Haematomas : extradural, subdural, and intracerebral, b) Aqueductal stenosis c) Intracranial abscess and granuloma d) Obstructive hydrocephalus. The symptoms of increased intracranial pressure include headache, drowsiness and blurring of vision. The headache is of moderate severity and can be associated with projectile vomiting. Since this vomiting is central in origin, there is no preceding nausea. Further increases in pressure causes changes in the level of consciousness with the patient becoming drowsy. The brainstem is compressed and pathologically shows small haemorrhages "duret" haemorrhages. Pupillary changes have been described previously, and will not be repeated here but remember an ipsilateral large pupil that does not react to light is seen.

Some intracranial tumours are associated with a change in personality. The patient becomes placid and withdrawn or hyperexcitable. The ability to concentrate suffers and there is loss of recent memory. Seizures may occur due to focal brain involvement. Partial seizures are common in gliomas and granulomas. Focal seizures that begin after the age of twenty are more likely to be due to tumours, in fact in some series the incidence has been reported to be as high as 10%. We shall now discuss the features of tumours in terms of their location.

TUMOURS OF THE FRONTAL LOBE

Loss of initiative, placidity and apathy may be the presenting features of a frontal lobe tumour. Sometimes the patient may display a rather silly sense of humour, with a tendency to use puns, the so called 'witzelsucht' that is due to loss of frontal inhibition. The patient may lose his job because of lack of interest, he may become unconcerned about his dress and personal appearance, and may even be euphoric. Seizures are commoner with frontal lobe tumours, and may be focal motor or *adversive* in type. An *adversive* seizure consists of head and eyes turning opposite to the site of discharge, and on some occasions progressing to a generalised seizure.

Except for dementia and an abnormal affect, there may be few physical signs elicited. A grasp reflex may be present. The examiner clasps the patient's hand and then lets go suddenly. If the grasp reflex is present the patient involuntarily grasps the examiner's fingers. A bilateral grasp reflex has no localising value, but a unilateral grasp reflex points to contralateral frontal lobe pathology. Tumours that infiltrate the motor strip lead to contralateral motor signs that vary from a mild weakness of the leg to a hemiparesis. A lesion that affects the speech area causes disturbances in speech. A parasagittal tumour causes bilateral leg weakness and sphincter symptoms due to involvement of the paracentral lobule. There may be mild focal weakness with some frontal tumours as the face area is more anterior than the arm and leg. False localising signs occur when a cerebellar tumour causes hydrocephalus and the frontal lobe is subject to increased pressure. Signs of frontal lobe involvement may then lead to faulty localisation. Tumours that involve the orbital surface of the frontal lobe may grow to a considerable size before causing symptoms.

TUMOURS OF THE PARIETAL LOBE

The parietal lobes contain the sensory cortex, hence tumours in this region cause sensory seizures that consist of paraesthesias in the opposite half of the body. Lesions of the dominant parietal lobe cause the "Gerstmann syndrome" with agraphia, finger agnosia, acalculia, astereognosis, and loss of two point discrimination. Non-dominant parietal lobe tumours lead to dressing apraxia, loss of body image and loss of topographical memory.

Sensory inattention is the inability to recognise stimuli on one half of the body. When both sides of the body are touched simultaneously

the patient cannot recognise stimuli on the affected side. Involvement of the visual association areas lead to visual inattention, with the patient tending to bump into objects in the affected visual field, and inability to perform motor action in the presence of normal strength, is also a feature of parietal lobe tumours.

TUMOURS OF THE OCCIPITAL LOBE

Tumours can destroy the visual cortex or affect the visual association areas. The more posterior the location of the tumour, the more simple the visual hallucination that may result in a seizure. Flashes of light, coloured lights and circles of light may be reported in the involved visual field. A hemianopia is a common defect in occipital lobe lesions, though altitudinal defects have also been observed. Sparing of macular vision with a hemianopia occurs when the tumour spares the area of macular representation, the occipital pole.

TUMOURS OF THE TEMPORAL LOBE

The temporal lobe is the most complex lobe of the brain. Rolled up in the manner of a scroll, encased in the narrow confines of the middle cranial fossa, it is as difficult for the student to visualise, as the other lobes are relatively simple. Not only does a region of the temporal lobe such as the hippocampus have different blood supply, but within the same area there is a different vulnerability to oxygen lack. The temporal lobe is also diverse in its functions, it plays a role in emotional, motor and autonomic activity. Therefore, it is not surprising that tumours of the temporal lobe may present in many different ways. Lesions that involve the anterior temporal lobe result in 'Psychomotor' seizures. Other temporal lesions may cause disturbances in cognition, or dysamnesic symptoms 'deja vu' and 'jamais vu'. 'Deja vu' is a feeling of familiarity with an unfamiliar situation, whilst 'jamais vu' is the sensation of feeling strange in familiar situations. Hallucinations of taste, smell, and vision may occur depending upon the site of tumour. Disturbances in affect such as spells of intense fear, or depression have been reported. Seizures that consist of automatisms are common, with the automatisms involving the face and especially the lips and tongue. If the Meyer's loop is involved a superior quadrantic field defect may be demonstrated. Problems with memory may be a prominent feature in lesions that involve the hippocampal region. Rarely sexual disturbances may occur.

TUMOURS OF THE BRAINSTEM

The brainstem consist of three parts : the midbrain, the pons, and the medulla. The structure of the brainstem will be reviewed before

discussing the tumours in this region. The corticospinal fibres descend via the cerebral peduncles and then through the basis pontis before decussating in the medulla. Fibres are so arranged in the midbrain that fibres meant for the leg lie most laterally, whilst fibres for the arm are located more medially. Fibres supplying the motor nuclei of various cranial nerves are the most medial in location. The sensory tracts unite in the midbrain and lie anterior to the nucleus of the third nerve. In the pons the medial lemniscus is located somewhat laterally in the corpus trapezoidium. The spinothalamic tracts lie posterolateral in relation to the medial lemniscus. In the medulla the gracile nucleus is medial and the cuneate nucleus lies just lateral to it. Nuclei of cranial nerves are generally located dorsal to the ascending and descending pathways.

The commonest tumour of the brainstem is a glioma, that occurs more frequently in children and is very slow growing. In India a tuberculoma is equally likely, causing some neurosurgeons to treat a brainstem 'tumour' with antituberculous therapy before considering a neoplastic lesion.

A brainstem glioma causes sixth and seventh nerve weakness, with the child showing signs of diplopia and facial weakness. Signs of pyramidal tract involvement such as hyperreflexia and spasticity are present, though frank paralysis occurs only very late in the disease. As the tumour grows ataxia may appear. Sensory signs are, however rare, and occur in only about a fifth of patients. Though vomiting may result from medullary involvement, headaches are uncommon because hydrocephalus does not occur till very late in the illness.

TUMOURS OF THE PITUITARY GLAND

These tumours lie in close proximity to the optic chiasm, and cause bitemporal or binasal hemianopia. Optic atrophy occurs frequently and papilloedema has also been reported. Disturbances of endocrine function like gigantism, acromegaly or Cushing's disease may occur. The most common pituitary tumour of childhood is the craniopharyngioma, whilst the chromophobe adenoma occurs in older individuals.

TUMOURS OF THE PINEAL BODY

"Parinaud's syndrome", a paralysis of upward gaze is found with tumours of the pineal body. A mild bilateral ptosis, dilated pupils and sluggish or even absent pupillary light reflex may be observed. Tumour in this region leads to rapid development of hydrocephalus and changes in the level of consciousness. Precocious puberty has been reported in pineal tumours.

TUMOURS OF THE THALAMUS AND BASAL GANGLIA

These tumours produce personality changes, and signs of extra-pyramidal involvement such as parkinsonism choreoathetosis and dystonia. Hemiparesis may be present due to involvement of the adjacent internal capsule.

TUMOURS OF THE EIGHTH NERVE

These tumours have already been described under the discussion of the eighth nerve (P. 55). A few additional features are described here. The acoustic neuroma comprises about 8% of all intracranial neoplasms and 80% of all tumours in the cerebellopontine angle. There is a 2:1 preponderance in females as compared to males. Symptoms of cranial nerve involvement occur early, increased intracranial pressure occurs very late in the disease.

TUMOURS OF THE CEREBELLUM

These tumours can occur at any age, but the medulloblastoma and astrocytoma are more common in childhood. Haemangiomas may form a part of the Phakomatosis. (Von Hippel Lindau's disease). Cerebellar tumours cause ataxia and in the case of medulloblastoma rapid signs of increased pressure with papilloedema and hydrocephalus. Separation of sutures occurs, and signs of cerebellar disease such as titubation, hypotonia, dysmetria, and nystagmus are seen.

COLLOID CYSTS OF THE THIRD VENTRICLE

These cysts are often mobile and can occlude the foramen of Monro or the aqueduct of Sylvius producing an acute hydrocephalus. Intermittent obstruction of the foramina result in severe paroxysmal headache which is relieved by a change in position of the head. These tumours can cause sudden death. Other signs of third ventricular involvement like somnolence, polyuria, obesity, loss of libido, are rare with colloid cysts but occur with third ventricular gliomas.

DIFFERENTIAL DIAGNOSIS OF AN INTRACRANIAL MASS

An intracranial abscess is the most important differential diagnosis that should be considered. Abscesses tend to be located in the temporal lobe and cerebellum when they occur due to direct spread of infection from the inner ear. Abscesses that are secondary to haematogenous spread are usually located in the parietal lobes. Frontal lobe abscesses are likely to be the result of extension of an infection in the frontal or

ethmoidal sinuses. The presentation of an abscess can be acute or chronic. Acute lesions cause mild fever with leucocytosis. Signs of cerebritis and meningeal irritation may be present. There may be a history of ear discharge, or sinus infection. Chronic abscesses may sometimes only be diagnosed at surgery. Papilloedema occurs late with abscesses but there may be widespread cerebral oedema.

Tuberculomas are a common cause of space occupying lesions in our country. In children they tend to be located in the posterior fossa. After the age of twenty they are usually supratentorial in location. Presence of tuberculosis elsewhere in the body, a positive Mantoux test and an elevated ESR should raise the suspicion of tuberculomas, though of course it is possible for a patient with tuberculosis to have a primary intracranial tumour. If the patient's visual deficit or clinical status is not grave enough to warrant immediate surgery, some neurosurgeons prefer to try antituberculous medication first, in suspected cases of 'tumour' in India. Fungal granulomas and syphilitic gummas are rare. Fungal lesions should be suspected in chronically ill and debilitated individuals and in immuno-suppressed patients.

Congenital lesions such as aqueductal stenosis, the Chiari malformation, and the Dandy-Walker syndrome may be mistaken for a tumour if the course is atypical or there is sudden decompensation. Hypertensive encephalopathy leads to a sudden change in the sensorium, focal neurological deficit, and seizures. The symptoms subside when the blood pressure is brought under control. Chronic subdural haematoma is another possibility, especially in older individuals with a history of gradual mental deterioration and periods of alertness and lucidity.

Pseudotumour cerebri is a diagnosis of exclusion, and is found to occur in young, obese females. There is a history of headaches, and blurring of vision. The blind spot is enlarged, there is bilateral papilloedema, but no focal neurological deficit. Sometimes there is a history of menstrual dysfunction, or of use of oral contraceptives. A cerebral venous sinus occlusion should be ruled out if there is a history of ear discharge or rarely of oral contraceptive use.

In India pseudotumour cerebri can be secondary to cysticercosis. Vitamin A intoxication, and polycythaemia are rare causes of pseudotumour cerebri. This condition is also known as "benign intracranial hypertension". All laboratory test including EEG, radionucleotide brain scans and C.T. scans are normal. Very small, slit like ventricles have been described with pseudotumour, but they can be seen in normal young people.

The presence of an intracranial neoplasm should be suspected in any patient who gives a progressive course, has focal findings, and partial seizures. The course is very short with malignant tumours like glioblastoma multiforme, but can be very gradual as in the case of acoustic neuromas and brainstem glioma.

INVESTIGATION OF A CASE OF INTRACRANIAL TUMOUR

They include invasive and non-invasive procedures. We shall first review the non-invasive tests and then describe the invasive procedures.

Skull X-rays Four views of the skull are obtained; antero-posterior, two lateral views (right and left), and a half-axial view (Towne's view). Further views may be required depending upon the site of the tumour. The Stenver's view may be necessary to judge the size of the internal auditory meatus in cases of acoustic neuroma. In children increased intracranial pressure secondary to hydrocephalus leads to a 'silver beaten appearance' with gyral markings being seen distinctly on the inner table of the skull. Separation of sutures may be present. Percussion of the skull may produce 'cracked pot' sound (Macewen's sign).

In adults the signs of increased pressure are different. Decalcification of the posterior clinoid process, and erosion of the dorsum sellae are seen. Meningiomas may produce local erosion of bone, and an increase in the markings of meningeal arteries. Calcification is another important sign of slower growing tumours, with calcification in meningiomas, gliomas, and pinealoma. Sellar calcification occurs with pituitary tumours.

The pineal gland normally does not calcify before the age of fourteen. Pineal calcification in older persons can help in demonstrating a shift of the brain substance. When the pineal calcification is deviated by a distance of more than 3 mm it is abnormal.

Electroencephalography The EEG may be helpful in localising the lesion. It is abnormal in about 70% of cases of hemispheric tumours and tends to be normal in tumours of the posterior fossa, sellar and hypothalamic region. Lesions that lie close to the cerebral cortex are more likely to produce focal abnormality than deep seated tumours. The EEG shows focal slowing in the area overlying the tumour. This slowing is in the delta range (1-3 Hertz.), and progressively increases with tumour growth.

Echoencephalography Transmission of ultrasound through the brain and its deflection by midline structures like the third ventricle may help in deciding whether or not there has been a shift of brain substance. This test is of limited value and is not used when other, more specific tests such as C.T. scanning and angiography are available.

Radio-isotope scanning It is positive in 70-80% of tumours but may be negative in sellar lesions and in avascular lesions. Its diagnostic yield is highest in lesions that have increased vascularity, such as meningiomas, arterio-venous malformations, abscesses, and metastasis. Lesions in the supratentorial compartment are more likely to be detected than lesions in the posterior fossa. A radio-active isotope (technitium) is injected intravenously, and the brain is scanned with a gamma camera. The isotope is concentrated in areas of increased vascularity or in regions of breakdown of the blood brain barrier. An acute cerebral infarction can be differentiated from a tumour by the following : Tumours will give a positive scan immediately after an acute event, whilst infarcts are generally negative initially and show the greatest positivity 7 days after occurrence. An infarct resolves, so that the abnormality decreases with time, and the scan is normal 3 months after the stroke, whilst a tumour shows an increase in the area of abnormality with passage of time. In some instances however, it is very difficult to differentiate between the two conditions.

Computerised tomography has revolutionised the investigation of intracranial pathology. At present it is not available in India except at a few major centres, hence its use is very restricted. It is the best investigation in terms of positivity, with over 95% of tumours being detected. It not only localises the tumour, but gives clues about tumour pathology, and also permits accurate judgement of the extent of oedema and shift of brain substance.

Cerebral angiography It is the mainstay of investigation in India, but its use will decrease drastically when C.T. scanning becomes widely available. Angiography is very useful in detecting shift of brain substance, and in the case of tumours shows a variety of abnormality. It can show abnormal vasculature, the so called 'tumour blush', it shows early draining veins that indicate local arteriovenous shunting, and it can show the tumour displacing and stretching major arteries.

Air encephalography It is used to study cases with suspected posterior fossa tumours, especially brainstem gliomas where arteriography has

failed to provide conclusive evidence. Sometimes ventriculography using contrast material is used to outline the lateral ventricles, and to analyse their configuration for distortions and changes in size. Both arteriography and airencephalography are done only when there is a high index of suspicion and when non-invasive tests are unhelpful in deciding management.

MANAGEMENT OF AN INTRACRANIAL TUMOUR

It consists of two parts: treating the increase in intracranial pressure, and treating the causative lesion.

Management of increased intracranial pressure Before we detail specific drugs that reduce intracranial pressure, we will review some concepts of the basic pathophysiology of cerebral oedema; Cerebral oedema is best defined as an increase in brain volume that is due to an increase in its water content. Brain oedema is of three types: Vasogenic, cytotoxic, and interstitial. Intracranial tumours are associated with vasogenic oedema. Vasogenic oedema is caused by an increase in capillary permeability, and is chiefly localised to the white matter. The oedema fluid consists of plasma filtrate including plasma proteins. This type of oedema responds to steroid therapy, and responds to osmotic agents only acutely. Acetazolamide and frusemide are of questionable value. Dexamethasone in doses of 12-16 mgm is given intravenously in divided doses and dramatically reduces the oedema. Mannitol is a very useful osmotic agent. 20% Mannitol given in a 300 ml solution over 2-3 hours reduces intracranial pressure on an acute basis. The longer duration over which the solution is administered reduces the rebound rise in intracranial pressure that often occurs when Mannitol is given rapidly. Sub-temporal decompression and ventriculoatrial shunt procedures are done in life threatening situations and in cases where cerebral oedema proves intractable to medical therapy.

Management of the tumour mass It depends on the nature and location of the tumour. Ideally the tumour should be resected in toto, but this may be impossible. Some tumours are very radiosensitive and respond well to radiation (medulloblastomas).

Meningiomas are generally fully excised when they are located in an accessible region. Gliomas are treated by a combination of surgery and radiation. The outlook for malignant neoplasms like glioblastomas is grim. Metastatic lesions are usually treated with radiation. A solitary metastatic lesion may be excised if the primary tumour has a high 5 year survival rate.

CHAPTER – X

TUMOURS OF THE SPINAL CORD.

By common usage the term spinal cord tumours implies tumours arising from within the cord, those arising from its roots, and from its meninges. Tumours of the vertebral column and other lesions that compress the cord like granuloma are excluded from this review.

The tumours may be classified by their location:

TABLE 10.1

1. EXTRADURAL

- a) **Metastasis** from a primary malignant neoplasm elsewhere in the body form the largest number of extradural tumours. The primary tumours involve the lungs, breast, lymphoid tissue and prostate. Often a metastatic tumour in the vertebral body causes it to collapse and compresses the spinal cord. Multiple myeloma may present in this manner.
- c) **Schwannoma** Though it occurs intradurally more frequently, both intra and extradural growth in the form of a “dumb-bell” may occur.
- c) **Meningioma** It is also common intradurally. Rarely an extradural variety has been seen.
- d) **Extradural sarcoma**
- e) **Lipoma**

INTRADURAL—EXTRAMEDULLARY here a tumour lies within the dura but outside the spinal cord.

- a) **Schwannoma**
 - b) **Meningioma**
- } These are the commonest tumours in this location.
- c) **Metastasis from a primary outside** the spinal canal and central nervous system.
 - d) **Metastasis from the central nervous system.** Tumours like medulloblastoma and ependymoma.
 - e) **Lipoma.**

3. **INTRAMEDULLARY** Tumours within the substance of the cord.
 - a) Glioma: either as ependymoma or astrocytoma. These are the commonest intramedullary tumours.
 - b) Haemangioblastoma
 - c) Schwannoma (a rare tumour at this site)
 - d) Metastasis (also rare at this site).
 - e) Lipoma - usually as part of spinal dysraphism (spina bifida)

Signs and symptoms The symptoms of cord compression differ depending upon the site of compression e.g. extradural, extramedullary or intramedullary, and according to its segmental level in the cord. The similarity of symptoms produced by the tumours at different locations, however rarely allows clinical differentiation of the site of cord compression.

Spinal compression affects the cord in two respects:

- 1) Direct pressure that interferes with conduction in the nerve roots and the spinal cord.
- 2) Compression of the spinal arteries, the radicular arteries and the veins leading to ischemia of the cord. It in turn leads to oedema and consequent degeneration of the ganglion cells with demyelination of the nerve roots and the tracts. Hence, it is imperative to detect cord compression early, so that decompression results in total recovery.

Symptoms Pain is the first symptom. In extradural and extramedullary tumours pain occurs in the distribution of the nerve roots at the level of the tumour. In the dorsal region, these pains are designated as "girdle pains" as the dorsal roots supply sensation to the trunk. These pains are intensified by coughing, sneezing and movements of the spine. Local pain in the spinal column may also be present and is most marked in the case of tumours of the cauda equina. Burning dysesthesiae and paraesthesias in the form of tingling occur late in the illness and tends to involve the distal parts of the lower limbs due to compression of the spino-thalamic tracts. Paraesthesias may occur early with extramedullary tumours.

Motor symptoms appear soon after as the compression progresses. The patients complain of stiffness of the lower limbs followed by

weakness of the muscles of the pelvic girdle. Weakness is first noticed on climbing stairs, or on getting up from a low position such as squatting. Walking long distances or running becomes difficult. Motor symptoms occur early in extradural and intramedullary tumours. The weakness progresses to a spastic paralysis with patients complaining of flexor spasms.

Sphincter disturbances develop very late in extramedullary lesions with hesitancy and retention of urine. Constipation develops in some patients and persists through out the course of the disease. Intramedullary tumours tend to involve the sphincters early as compared to extramedullary tumours.

The signs of cord compression are discussed under motor signs, sensory signs, local and autonomic changes :—

Motor

- I) Early involvement of the the anterior root especially in the cervical and lumbar region, leads to weakness and wasting of the involved muscles which also show fasciculations.
- II) Compression of the corticospinal tracts leads to spasticity and weakness in the lower limbs. As the disease progresses and the compression intensifies, total paraplegia may develop. Initially there is paraplegia in extension, followed by paraplegia in flexion. In a high cervicomedullary junction compression (cranio-vertebral anomalies), weakness first appears in one upper extremity, followed by a clockwise or anticlockwise progression of weakness in the other three extremities, i.e if weakness first appears in the left arm, it is followed by weakness in left leg then the right leg and finally the right arm.

Sensory

- I) If the posterior roots are compressed hyperaesthesia and hyperalgesia occur in the distribution of the involved nerve root. Later analgesia and anaesthesia are noted.
- II) Compression of the spinothalamic tracts leads to loss of pain and temperature sensations below and opposite to the side of the lesion. Touch is preserved as it has double pathways in the cords "dissociated anaesthesia".
- III) Impairment of joint position sense and vibration sense occurs early if the compression is located posteriorly.

Brown-Sequard syndrome (Hemisection of the cord)

There is loss of pain and temperature sensations on the side opposite to the lesion. Weakness and spasticity occurs on the same side as the lesion. Therefore, a right sided lesion, will result in sensory disturbance on the left and the motor paralysis on the right.

Local signs

- I) Tenderness of the spine may be demonstrated in extra and intra-dural tumours but is rare in intramedullary lesions.
- II) When patient flexes the neck, a wave of paraesthesia descends through the body (Lhermitte's sign). It may be present in any lesion that involves the posterior columns and has been noted in cervical spondylotic myelopathy.
- III) Spasm of the spinal muscles, may result in limitation of movements of the spine, at the level of the lesion.
- IV) Spinal scoliosis may be noted if the spasm is unilateral.

Reflexes

The reflex arc subserved by the root or the segment involved is broken. Hence, that particular tendon reflex is absent, while the reflexes below the level of the lesion are usually exaggerated. The plantar reflexes are extensor and the cremasteric and abdominal reflexes are diminished or absent. If the C6 root or segment is involved, the biceps jerk is absent but triceps jerk and the lower limb reflexes are exaggerated. Inversion of the reflex occurs due to overflow phenomenon in the affected spinal segment. In case of biceps jerk, the overflow involves the flexors of the fingers or the triceps, so that on trying to elicit the biceps tendon reflex, there is no contraction of the biceps but the fingers will still flex or the arm may extend because of contraction of the triceps.

Autonomic changes

These are of poor localising value. However, excessive sweating may occur below the level of lesion. It is important to localize cord compression at various levels of the cord, i.e. cervical, thoracic, lumbar and sacral segments. The principles that govern clinical localisation are detailed below.

General principles

At the level of the lesion, the anterior horn cells or the root will be damaged giving signs and symptoms of lower motor neurone involvement,

in the form of wasting, weakness and fasciculations. For instance involvement of the first dorsal level gives weakness and wasting of the small muscles of the hand, whilst a lesion of the fifth lumbar segment results in a foot drop. Thoracic level involvement may go unnoticed by the patient and can only be spotted by a dilligent examiner. Intercostal weakness may be observed. Appropriate tendon reflexes are absent so that a lesion at fifth cervical level will cause absence of the biceps jerk.

Upper motor neurone involvement produces a spastic paraplegia, leading to symptoms of leg stiffness and an inability to walk on uneven ground. With increasing spasticity the gait becomes markedly stiff. Examination at an early stage may reveal only a minimal increase in tone with brisk reflexes. The abdominal reflexes may be absent the plantar response extensor. As the lesion increases in size the spasticity becomes more marked, and patellar or ankle clonus may be elicited. Weakness due to a pyramidal lesion usually appears first in the extensor group of muscles in the upper extremity and the flexor group of muscles in the lower extremities. Therefore, shoulder abduction, elbow extension, wrist extension and finger abduction will be weak in the arm, while in the legs hip flexion, knee flexion and dorsiflexion of the foot is affected.

Pain results from nerve root involvement and may be dull aching, shooting or knife-like. At times hyperaesthesia occurs and the patient may describe the area as 'sore', or as if "a sand paper has been rubbed on it". The pain generally extends from the back along a given dermatome (girdle pains in dorsal region).

Damage to the spino-thalamic tracts and dorsal columns produce symptoms of tingling, or numbness. Deep dull aching pains or shearing or cold feeling result from spino-thalamic involvement. Patients may state that they get a feeling as if they are walking on cotton wool. The pain and temperature sensations are diminished or absent below the level of the lesion. Touch sensation is usually preserved as it has dual pathways. Vibration sense also may be impaired below the level of lesion. Similarly, the joint position sense at the toes may be impaired if the dorsal columns are affected.

We shall now briefly review features of individual segmental involvement at various levels.

CERVICAL CORD LESION

Cervical 1 to 4 Patients will complain of upper nuchal pain with stiffness of the neck. The pain may extend to the supra-clavicular region

in C4 lesions. A spastic quadruparesis with a clockwise spread of weakness and a sensory level extending upto clavicle and involving both upper extremities is seen.

Cervical 5 and 6 Pain and loss of sensation occurs along lateral border of the arm in (C5) and the forearm to the thumb in C6 lesions. The biceps and the supinator reflexes are absent or inverted. The deltoid, supraspinatus, infraspinatus, rhomboids, biceps, and brachioradialis muscles are weak. The triceps, knee and ankle jerks are exaggerated. The abdominal and cremasteric reflexes are absent and the plantar responses are extensor. The sensory level may be anywhere below the clavicle depending on the damage caused to the pathways by the evolving lesion. In a fully evolved lesion the sensory level is at the clavicular region.

Cervical 7,8 and dorsal levels Pain and loss of sensation will extend from axilla (D1) to the triceps (C7) and the medial forearm region including the little and ring fingers (C8). The triceps jerk will be absent and the biceps and supinator jerks will be normal. The other signs and symptoms are similar to a C5,6 lesion.

Dorsal 2 to dorsal 7 Pain and hyperaesthesia or loss of sensation occurs in the region of the dermatome involved (Girdle pains). The sensory level will be three segments below the level of lesion, that is if the fourth dorsal segment is involved, the sensory level will be anywhere below the D7 level. Spastic paraplegia will be present.

Dorsal 8 to 10 and 10 to 12 Pain and loss of sensation will depend upon the dermatome involved, but weakness can be demonstrated in the upper abdominal muscles. if the examiner asks the patient to lift the head and trunk off the bed, the umbilicus is dragged downwards, due to non contraction of upper abdominal and normal contraction of lower abdominal muscles. In D 10 to D 12 lesions the reverse occurs and the umbilicus moves upwards. The sensory level and pyramidal involvement follow the pattern of upper thoracic level tumours.

Conus medullaris and cauda equina lesions The common lesions of the conus medullaris are ependymoma or spinal dysraphism (a dermoid or lipoma). The lower sacral roots are involved leading to pain in the perianal and genital regions. The sensations in these areas is diminished, and the patient may complain of impotency. Lowback pain is common and dribbling incontinence almost always occurs. The anal reflex is

absent, the knee jerks are brisk but the ankle jerks may be lost, with a foot drop. If the extensors of the toe are spared, a Babinski response may be elicited.

Cauda equina lesions may be either lateral or medial. The usual cause of a *lateral* lesion is neurofibroma and rarely a high lumbar disc prolapse. Pain in the anterior thigh, with diminution of sensation in the distribution of L 2 or L3 segments is noted. The quadriceps muscle is weak and knee jerk is absent due to L2,3 roots being involved. If there is pressure on the conus, the ankle jerk may be brisk with an extensor plantar response. A large disc extrusion may even cause sphincter incontinence.

Midline or medial cauda equina lesions lead to bilateral lumbar and sacral root involvement with pain along both sciatic nerves. Weakness and wasting of the muscles of the feet along with involvement of the hamstrings and glutei is seen. The knee jerk may be preserved but the ankle jerk is absent. Sensory loss involves appropriate dermatomes, depending upon the roots involved. Tumours that lead to this type of lesion are chordomas, metastatic tumours, leukaemic infiltrates or seeding from medulloblastoma.

The clinical diagnosis is confirmed by the following investigations:

- A) **Plain roentgenograms of the spine** changes produced by spinal cord tumours include :
 - i) Thinning of the pedicle with erosion and disappearance.
 - ii) Enlargement of an intervertebral foramen-(common in schwannomas).
 - iii) Erosion of other parts of the vertebra (lamina, bodies, junction of the two laminae in cervical vertebra)
 - iv) Widening of the antero-posterior and lateral diameters of the bony canal. This is seen in intramedullary tumours of childhood, and in syringomyelia.
 - v) Rarely a collapsed vertebral body may be seen. (multiple myeloma).
- B) **CSF examination** It is not diagnostic, but may indicate the possibility of a spinal cord tumour. The following changes may be observed.
 - i) Rise in CSF protein.
 - ii) Xanthochromia

- iii) Queckenstedt's test: Failure of rise in CSF pressure on jugular venous compression indicates a subarachnoid block. It is not very useful due to many false negative results, and is rarely used today.

If a lumbar puncture is performed a few days before myelography, it makes myelography difficult and risky due to a leak of CSF from the theca and collection of CSF outside the subarachnoid space. Myodil may not be introduced in the subarachnoid space. CSF should therefore be obtained at the time of myelography.

Myelography It is the examination of the spinal subarachnoid space by roentgenography after injection of contrast material (Myodil, Pantopaque), an iodinated compound-Iodophenindate. Other contrast materials used are Dimer-X, Amipaque or air. The patient is placed on a roentgenographic table that can be tilted. The contrast is then made to flow up when injected by the lumbar route by tilting the table so that the head is lowered. The contrast is either arrested or slowed down in the region of the tumour and at this juncture the roentgenograms are taken. Extradural, intramedullary and extramedullary lesion give characteristic pictures and it is usually easy to differentiate between them. In intramedullary lesions the classical 'wine glass appearance' of the dye column is seen at the level of the lesion.

Computerized Axial Tomography (CAT scan) of the spinal column is being utilized to a greater extent as an aid to diagnosis but its use is limited by its non-availability in all centres of our country.

Treatment Spinal tumours, by virtue of their causing compression of the spinal cord can only be treated surgically, by laminectomy at the appropriate level. Schwannoma are usually easy to excise and so are meningiomas. For removal of the schwannoma, the nerve root from which it is arising has to be sacrificed. Gliomas which are usually intramedullary, have defied successful surgical or medical treatment. With the use of the surgical microscope, ependymomas are easier to excise than gliomas, as the former has a good plane of cleavage, which allows a complete excision without causing much damage to the spinal cord. Gliomas are difficult to excise as the plane of cleavage is poor. Both these lesion are subjected to deep X-ray therapy after surgery. Haemangioblastoma though intramedullary is usually removable. Both short term and delayed results are good. Metastasis are usually extradural and invariable present with rapid onset of paraplegia. They are treated

by immediate laminectomy, removal of as much tumour as possible, followed by deep X-rays and chemotherapy. The results are generally not good. Metastasis from primary brain tumours specially medulloblastoma are not very common, but when present are multiple and wide-spread. These are usually treated by radiotherapy and chemotherapy. Lipomas form a part of congenital spinabifida and are in a younger age group. The intramedullary and extramedullary varieties usually cause cord compression. These are intimately adherent to the cord and to the nerve roots and cannot be easily excised for fear of injury to the cord. They are therefore, treated with a decompressive laminectomy and opening of the dura. This procedure creates more room for the cord, thus decreasing the severity of compression.

The tables below correlate spinal segments with clinical features and differentiate between extradural, intradural and intramedullary tumours.

CORRELATION OF SPINAL SEGMENTS WITH CLINICAL FEATURES

Level of lesion	Symptoms & signs	Muscles affected	Reflexes
Above C5	Nuchal pain, quadraplegia.	Trapezius, pectorals	All reflexes brisk
C5—6	Pain and loss of sensation in the lateral border of arm and forearm. paraplegia, sensory level below clavicle.	Deltoids, biceps, brachio-radialis supra & infra spinati	Biceps and supinator lost or inverted other reflexes very brisk.
C7—T 1	Pain and loss of sensation from axilla to triceps, medial forearm and ring & little finger, paraplegia, sensory level below clavicle	Triceps, small muscles of hand	Biceps & supinator normal, triceps lost, knee & ankle brisk.

D2—7	Girdle pains, sensory, level 3 segments below level of lesion paraplegia.	Intercostals	Knee&anklebrisk
D8—10	Same as above	Upper abdominals weak.	Loss of upper abdominal reflexes KJ & AJ brisk.
D10—12	Same as above	Lower abdominals weak.	Loss of lower abdominals reflexes KJ & AJ brisk.
L2—3	Inguinal pain, minimal spasticity	Weakness of Quadriceps.	Knee jerk lost
Conus medullaris	Saddle anaesthesia sphincter incontinence	Sphincter muscles	Ankle jerk absent
Cauda equina	Radicular pain incontinence rare.	Asymmetric, foot muscles, hamstrings, glutei	Knee ankle jerk may be absent.

DIFFERENTIATION BETWEEN EXTRADURAL, AND INTRAMEDULLARY TUMOURS.

	Extradural	Intradural	Intramedullary
Pain-root distribution.	+	+	—
Pain local	+	±	—
Paraesthesae	early	early	—
Sensory loss	Rising level	rising level	Dissociated loss
Weakness	First	Later than Sensory	Early

Faciculations & Atropity	$\pm$	$\pm$	—
Sphincter	Late	Late	Early.
Trophic changes	$\pm$	$\pm$	+

CHAPTER — XI

INJURIES

HEAD INJURIES

Head injury is defined as any injury that results from a blow to the head with or without alteration of consciousness. Facial injuries that cause disturbances in consciousness are also included. Head injury is classified further into 5 categories :

- 1) **Trivial injuries** without loss of consciousness or post traumatic amnesia.
- 2) **Mild injuries** where the loss of consciousness / post traumatic amnesia does not exceed 30 minutes, and there is no skull fracture.
- 3) **Moderate injuries** where loss of consciousness / post traumatic amnesia lasts for more than thirty minutes but for less than 24 hours, and / or a skull fracture is present.
- 4) **Severe injuries** Where loss of consciousness / post traumatic amnesia lasts for more than 24 hours, and / or a cerebral contusion, and / or a intracranial haematoma was present.
- 5) **Fatal injuries** patient died following head injuries.

Categories 3, 4, 5, are generally accepted as evidence of structural brain damage.

Post traumatic amnesia is the duration for which there is a defect in continuous memory following a head injury. Anterograde amnesia is amnesia for events after the injury Retrograde amnesia is amnesia for events prior to the injury. It is very brief in most cases of head injury, and shortens with recovery even in very severe cases.

Cerebral contusion is a 'bruise' of cerebral tissue following head injury. Pathologically there is microhaemorrhage, with loss of neurones and resultant gliosis. Unlike a cerebral infarction the overlying arachnoid undergoes fibrosis as the injury involves the structures from the periphery to the centre. An infarct on the other hand involves areas within a common vascular system, thus sparing the meninges with their external carotid source of blood supply. The term 'concussion' generally means altered consciousness for a brief period following a mild head injury, but with amnesia for the event itself. However, it is used rather

loosely and has become synonymous with any head injury. We therefore prefer not to use the term, and instead classify head injuries according to the 5 categories described above.

INTRACRANIAL HAEMATOMA These include: epidural, subdural, and intracerebral. These haematomas are secondary to a localised collection of blood following head injury. When there is collection of CSF instead, the term hygroma is used.

EPIDURAL HAEMORRHAGE It is extradural collection of blood generally of arterial origin, usually located in the temporal region due to a tear in the middle meningeal artery. It occurs in about 1% of an unselected series of head injuries. It is rare under the age of two because the dura is tightly adherent to the inner table of the skull. A 'lucid interval' after the injury is more often absent than present. A fracture of the temporal bone is invariably present, and there may be blood behind the tympanic membrane (Battle's sign). The patient deteriorates rapidly, and constitutes a neurosurgical emergency. An angiogram or a C.T. scan confirms the diagnosis and subtemporal decompression with evacuation of the clot offers the only chance of saving the patient.

SUBDURAL HAEMATOMA These are further subdivided into :

a) Acute occurring in the first 3 days after trauma. They are most common in the first 24-36 hours after head injury. The injury is severe with a skull fracture present in over 80% of cases. The patient shows an increase in focal neurologic signs with deterioration in consciousness. Focal seizures may occur. There is generally underlying brain contusion and even presence of brainstem damage. The treatment of choice is evacuation by a craniotomy. The morbidity and mortality of an acute subdural is very high. The mortality is nearly 70% for patients over the age of 65, and few patients are able to lead an independent existence after suffering this severe head injury.

b) Subacute These cases are defined by the existence of a membrane which is a sprouting of fibroblasts from the inner layer of the dura. It is otherwise similar to an acute subdural, but has a slightly better outcome. It has been defined as a haematoma that becomes manifest between 4 and 20 days after a head injury.

c) Chronic Unlike the acute subdural these haematomas are found weeks to months after trauma. The initial injury may be so

trivial, hence forgotten. A third of the patients with a chronic subdural haematoma do not give any history of trauma. The bleeding is said to be venous in origin, with a crescentic shaped clot that compresses the underlying brain. The patient presents with impairment of memory, and may even be admitted to the psychiatric unit as a case of 'organic brain syndrome'. The mental status is abnormal with the patient mildly confused and disoriented. There are no focal neurological findings, and the diagnosis may be missed if not suspected. Some patient may complain of headache that is worse in the morning and aggravated by coughing or stooping. Seizures are uncommon.

The diagnosis is made by angiography or on C.T. scan. The EEG may also be abnormal showing unilateral slowing over the involved hemisphere. The haematoma is evacuated by a burr-hole, and is followed by a good recovery of cognitive function.

The cause of head injury depends on the environment of the patient. In highly industrialised countries automobile accidents are the commonest cause, though in the urban United States, the inner city 'ghettoes' show a higher incidence of interpersonal injury. Falls, and recreational injuries are other causes, whilst coconuts falling and striking individuals has been reported in south India.

The clinical evaluation of a head injury patient involves ascertaining the occurrence of loss of consciousness and post traumatic amnesia. Alcohol use in relation to head injury should be noted. Presence of other injuries must be noted, and skull x-rays done if there is alteration in consciousness after injury. Any patient with a vague history, or with altered mental status, or who complains of sleepiness should be admitted for a 24 hour observation period. Patients with rapid deterioration of consciousness and/or focal neurological signs should be evaluated by a neurosurgeon as soon as possible.

It is imperative that no drugs be used to dilate the pupils, since it distorts a very important sign of increasing intracranial pressure. Drugs that depress consciousness should not be used.

The management depends upon the extent of the trauma and the presence of fractures and intracranial haematomas. Depressed fractures may require surgery and haematomas have to be evacuated surgically as has been discussed in the previous section. Monitoring the Glasgow Coma Scale on a hourly or daily basis and treating increases in

intracranial pressure by osmotic agents are some of the other measures used in the care of head injured patients. Severely injured patients need long term rehabilitation and may be left with substantial cognitive and motor deficits.

INJURIES AT SPECIAL SITES

1) Hypothalamic injuries These produce hyperthermia, with the temperature rising above 42 degrees celsius. Erosion of the gastric mucosa leading to haematemesis, and affection of the hypothalamic nuclei leading to diabetes insipidus also occur.

2) Brainstem injuries These are common as a secondary feature after severe head injury. Coma, cranial nerve palsies and a poor prognosis are hallmarks of brainstem involvement.

3) Temporal lobe contusion There are 3 sites in the brain that have a predilection for developing cerebral contusion after a head injury. They are a) the frontal pole, the temporal pole and the occipital pole. These are regions where the brain slides and dashes against the uneven bony surface of the anterior and middle cranial fossae. A temporal lobe contusion can lead to signs of increased intracranial pressure as the oedema develops in that lobe. Some patients may be dysphasic, others become drowsy and develop unilateral cranial nerve palsies affecting the third, and sixth nerves. The diagnosis of temporal lobe contusion is made at angiography or on C.T. scan. The temporal lobe has to be decompressed surgically and the contused brain excised.

SPINAL CORD INJURIES

Any injury that is severe enough to cause damage to the spinal cord invariably injures the vertebrae, and the soft tissues such as the ligaments, and the muscles. Conversely if the bone and soft tissue are able to withstand the injury the spinal cord escapes injury.

Soft tissue injuries The muscles and ligaments can be injured in hyperextension and hyperflexion as occurs in sudden acceleration, or deceleration. Hyperextension is produced in acceleration injuries as when a car is hit from the back by another fast moving vehicle. A head on collision on the other hand produces sudden deceleration and results in a hyperflexion injury. The symptoms consist of pain at the site of the injury, and limitation of neck movement. There is usually no associated spinal cord injury. Rarely, hyperextension of the neck leads to a

paralysis where the arms are involved to a greater extent than the legs, sensory changes are minimal, and sphincter incontinence may be present.

Treatment of the common 'whiplash' syndrome consists of analgesics and rest with local application of heat and other palliative measures.

Bone injuries Dislocation of the vertebrae without fracturing the bone is only possible in the cervical region. The common sites of dislocation are the atlanto-axial joint, and dislocations of C4 over C5 and C6. Atlanto-axial dislocation can cause sudden death due to respiratory parralysis. If the patient survives he may be left with a residual quadraplegia, with loss of pain and temperature sensation upto the clavicular level, and with marked loss of joint and position sense in the arms. Dislocations at lower levels occur following falls whilst riding or diving into shallow waters. These actions cause excessive flexion of the spine with dislocation and result in compression of spinal cord between the lamina of the upper vertebra and the body of the lower vertebra. Quadraplegia with a upper thoracic level and with absence of the biceps and supinator reflexes may be found. Fracture dislocation is common at the level of the twelfth thoracic and the first lumbar vertebra. Fractures of the spinous and transverse processes and fissures or compression fractures of the lower thoracic vertebral bodies do not lead to neurological complications. Occasionally haematomyelia or extra dural haematoma may occur. The extradural haematoma must be evacuated at once.

Spinal concussion A disturbance of the synaptic transmission in the central gray matter results in total paralysis below the level of the lesion, hypotonia, loss of tendon reflexes, loss of pain and temperature and sphincter incontinence. There is preservation of fine touch and proprioceptive sensation as the posterior columns have no synaptic connections in the spinal cord. There is total recovery of function in 24-48 hours.

Spinal contusion It occurs due to nipping of the spinal cord between the lamina of the vertebral body above the lesion and the posterior edge of the vertebral body below the lesion. Petechial hamorrhages are widespread in the area. Oedema increases and haematomyelia may occur. The clinical features mimic those of a concussion of the cord except that the posterior columns are also involved. The sensory level ascends upwards due to increasing oedema. Recovery is poor and spasticity develops

after recovery from spinal shock. Haematomyelia resembles syringomyelia except that the symptomatology is acute in onset.

Treatment Injured nervous tissue does not regenerate. Though certain experimental measures like implantation of peripheral nerve in the injured region have been tried, there is no effective therapy for cord injuries. The aim of therapy is to prevent further injury and to attempt to realign the vertebral column adequately. Steroids are used to decrease oedema and surgery to remove impinged cartilage or bone and removing extruded discs, may be of value. Nursing care and rehabilitative measures are very important in severely injured cases and play a crucial role in eventual outcome.

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